Chapter 28
Ventricular Septal Defects

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28.1 Clinical Indications

The defects that may be suitable for percutaneous closure are located within the muscular septum (muscular ventricular septal defects, MVSD) or in the perimembranous septum (perimembranous ventricular septal defects, PVSD) with or without aneurism, and they can be native of residual post surgery.

Surgical repair is currently the only option for doubly committed or supracristal defects, for perimembranous defects associated with prolapse of aortic valve and aortic regurgitation and for any defect associated with malalignment of the muscular outlet septum or straddling and overriding atrioventricular valves.

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G. Butera et al. (eds.), *Cardiac Catheterization for Congenital Heart Disease: From Fetal Life to Adulthood*,
DOI 10.1007/978-88-470-5681-7_28, © Springer-Verlag Italia 2015

Large defects give signs and symptoms of cardiac failure in early infancy, and they have to be treated surgically during the first months of life.

Clinical indications for the closure of ventricular septal defects (VSD) are:

Symptoms of heart failure

Signs of left heart volume overload with an echocardiographic evidence of a significant left-to-right shunt through a VSD

A shunt is considered significant when the following are found:

(i) Left atrial enlargement, defined as a left atrial-to-aortic ratio >1.5
(ii) Left ventricular enlargement (left ventricular overload), defined as a left ventricular end-diastolic diameter >+2 standard deviation (SD) above the mean for the patient's age

Closure may be needed in order to prevent pulmonary arterial hypertension, ventricular dilation, arrhythmias, aortic regurgitation and development of double-chambered right ventricle. In specific cases small defects, with neither symptoms of cardiac failure nor overload, may need closure if an episode of infective endocarditis was experienced.

28.2 Patient Selection

- Absence of active infection: if a source of potential infection is found, treat it before catheterization.
- Complete and deep analysis of previous medical history, cardiac catheterization and surgeries if the VSD is a residual post-surgical defect.
- Check personally the TTE before start of the procedure.

- Take into consideration the possibility to treat associated anomalies if they are present (pulmonary valve or branch stenosis, atrial septal defect, etc.)
- Take personally the informed consent for all the planning procedures.

28.3 Technical and Equipment Issues

28.3.1 Device for MVSD

The Amplatzer muscular ventricular septal defect occluder (Amplatzer Muscular VSD Occluder, AGA Medical Corporation, St. Jude, MN) is a self-expandable device made of nitinol wires (thickness 0.004–0.005 in.), consisting of two flat discs having a diameter 8 mm larger than a central connecting waist (7-mm long) (Fig. 28.1). The diameter of the waist determines the size of the device, and it is available in sizes from 4 to 18 mm. Three Dacron polyester patches are sewn with polyester thread into both discs and the connecting waist. The device is secured to a delivery cable and is inserted into a delivery sheath ranging from 6 to 9 French in size.

28.3.2 Device for PMVSD

The Amplatzer membranous ventricular septal defect occluder has two discs of unequal size (Amplatzer Membranous VSD Occluder, AGA Medical Corporation, St. Jude, MN). The aortic rim of the asymmetric left ventricular disc exceeds the dimensions of the connecting waist by only 0.5 mm, so as to avoid impingement on the aortic valve, whereas the apical end

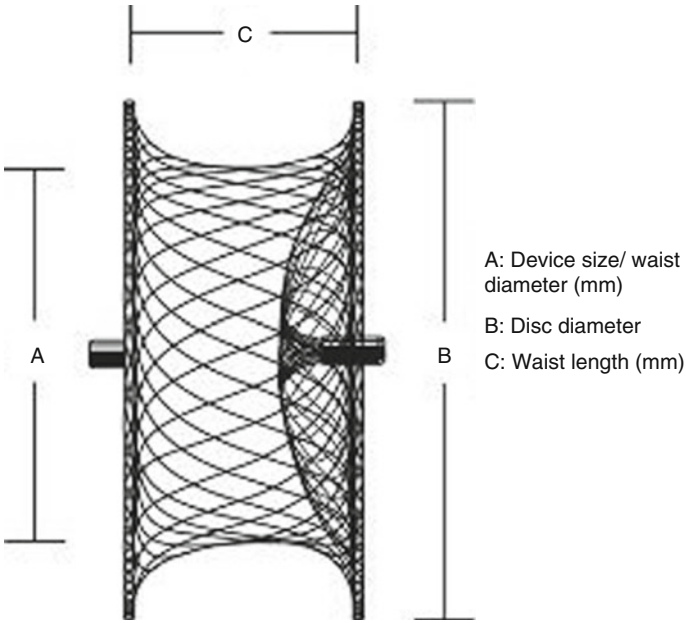

Fig. 28.1 The Amplatzer Muscular VSD Occluder, AGA Medical Corporation, St. Jude, MN. *A* device size/waist diameter (mm), *B* disc diameter (mm), *C* waist length (mm)

is 5.5 mm larger than the waist. This apical end of the left ventricular disc contains a platinum marker to facilitate correct orientation during implantation. The right ventricular disc is symmetrical, and it exceeds the diameter of the connecting waist by 2 mm throughout its circumference (Fig. 28.2). The device is available in sizes from 4 to 18 mm and requires delivery sheaths from 7 to 9 French.

A MVSD Amplatzer device II is also available. It was redesigned to prevent conduction abnormalities with a 75 % reduction in radial force, 45 % reduction in clamping force and 10 % increase in stability. The left disc of this new device has an

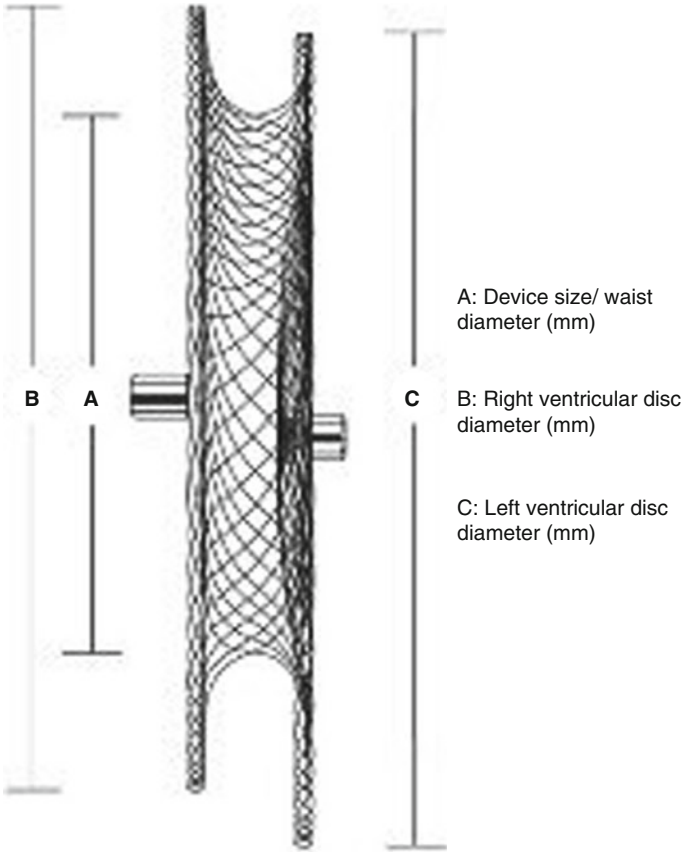

Fig. 28.2 The Amplatzer Membranous VSD Occluder, AGA Medical Corporation, St. Jude, MN. *A* device size/waist diameter (mm), *B* right ventricular disc diameter (mm), *C* left ventricular disc diameter (mm)

elliptical and concave shape that adapts to the LV outflow tract. It is available in two configurations:

1. Eccentric with a 1-mm superior rim and a 3-mm inferior rim
2. Concentric with a 3-mm superior and inferior rims

The waist length is increased from 1.5 to 3 mm, and the waist diameter ranges from 4 to 10 mm in 1-mm increments plus 12 and 14 mm. Polyester patches are sewn into the discs.

The delivery system consists of a delivery cable and a pusher catheter having a sharp curvature of 180° inferiorly. This allows correct orientation of the left ventricular disc during implantation. It has a flattened part of the socket that matches the flat portion of the microscrew, in order to force larger part of the left ventricular disc to be oriented downwards so that it points to the left ventricular apex.

28.4 Procedure

28.4.1 Preparation

- General anaesthesia and orotracheal intubation.
- Biplane catheterization laboratory preferred.
- Patient position with arms lifted up, behind patient's neck (attention to brachial plexus overstretching).
- Patient is fully monitored including an arterial line for continuous arterial pressure monitoring, two peripheral venous lines or a central venous line and vesical catheter for diuresis evaluation.
- A transoesophageal echocardiography 2D (2D-TEE) or 3D (3D-TEE) must be used to monitor the procedure.
- Heparinization with IV administration of 100 UI/kg heparin. Check hourly the activated clotting time >250 s. In case

administer heparin intravenously during the procedure. Usually it is not needed.
- Antibiotics IV: usually a cephalosporin.
- The procedure has to be considered as a surgical intervention. Therefore, special care has to be paid to strict asepsis. Special attention has to be given to operators' scrubbing and patient's preparation (including careful depilation). The personnel involved has to wear masks and hats.

28.5 Access Site

- A femoral vein (FV) access is used to approach the closure of a PMVSD, and an internal jugular vein (IJV) can be used for MVSD closure.
- An arteriovenous circuit must be created (internal jugular vein-femoral artery for MVSD and femoral vein-femoral artery for PMVSD closure).
- Both sides for vascular femoral access are prepared.

28.6 Catheterization and Haemodynamic Evaluation for MVSD Closure

Left ventricular angiographies are obtained in axial projections for best evaluation of VSD size and position, in addition to TEE views. Left ventriculography in the hepatoclavicular projection (35° left anterior oblique/35° cranial) is performed to imaging mid-muscular, apical posterior defects. Anterior defects are better seen in 60° left anterior oblique/20° cranial (Fig. 28.3).

The VSD is crossed from the left side by using a right Judkins catheter and a soft Glidewire (0.035″, J Tip, Terumo); the wire is advanced to the pulmonary artery or to the SVC/IVC,

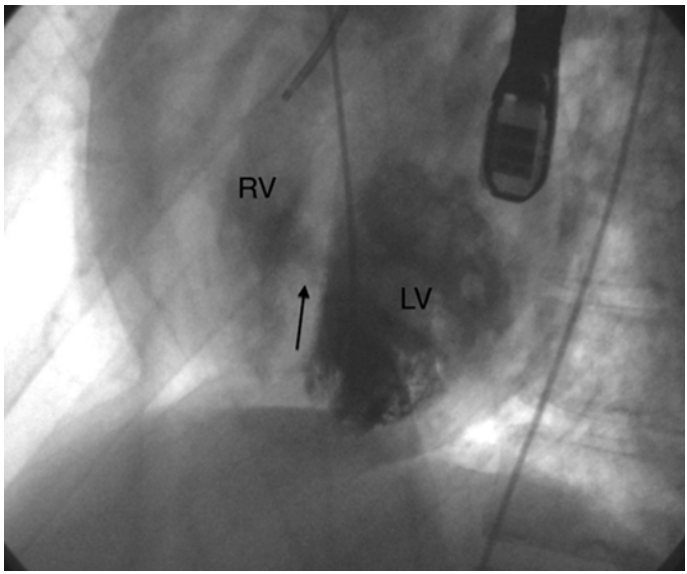

Fig. 28.3 Left ventricular angiogram: *LV* left ventricle, *RV* right ventricle, *Arrow* mid muscular VSD

where it is snared with a GooseNeck Snare (Microvena Corporation, 20–25 mm in adults, 10–15 mm in children) and exteriorized out of the right internal jugular vein or femoral vein establishing an arteriovenous circuit (Fig. 28.4).

Over the circuit, an appropriate size delivery sheath is advanced from the vein all the way until the tip of the sheath is in the ascending aorta. The dilator is withdrawn, and the sheath is pulled back in the left ventricle.

When the tip of the sheath is placed in the mid cavity of the left ventricle, the dilator and the wire are gently removed; a left ventriculogram is usually repeated, to confirm the position of the long sheath and also to obtain additional information on the position and the size of the VSD.

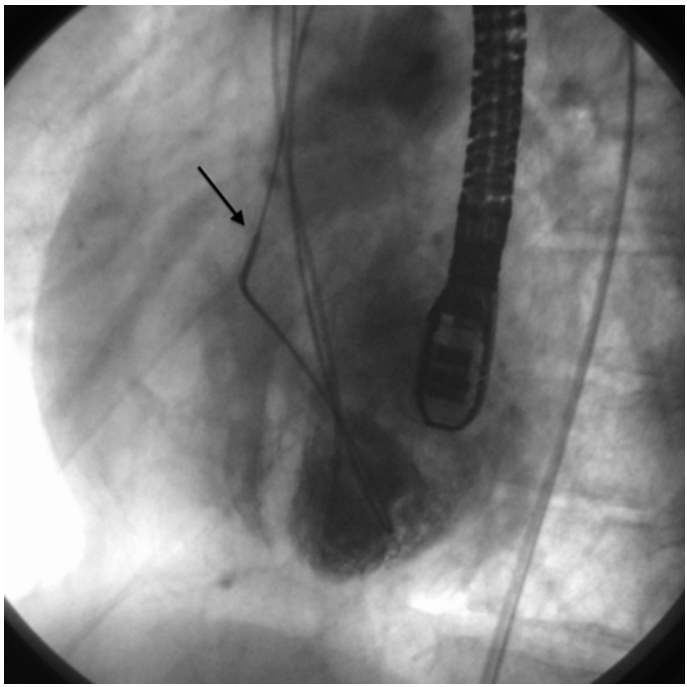

Fig. 28.4 Left ventricular angiogram: the *arrow* shows the arteriovenous circuit (femoral artery-internal jugular vein)

According to both angiographic and echocardiographic information, a muscular VSD occluder 1–2 mm larger than the maximum size of the defect is chosen.

The device is attached to the delivery cable, loaded into the plastic loader, introduced and advanced into the sheath.

The left disc is deployed in the left ventricular cavity, making sure it is not impinged in mitral valve apparatus, then the entire system is withdrawn towards the septum (Fig. 28.5a), and the central waist and the proximal disc are deployed; a test angiogram is done to verify the correct position of the device (Fig. 28.5b).

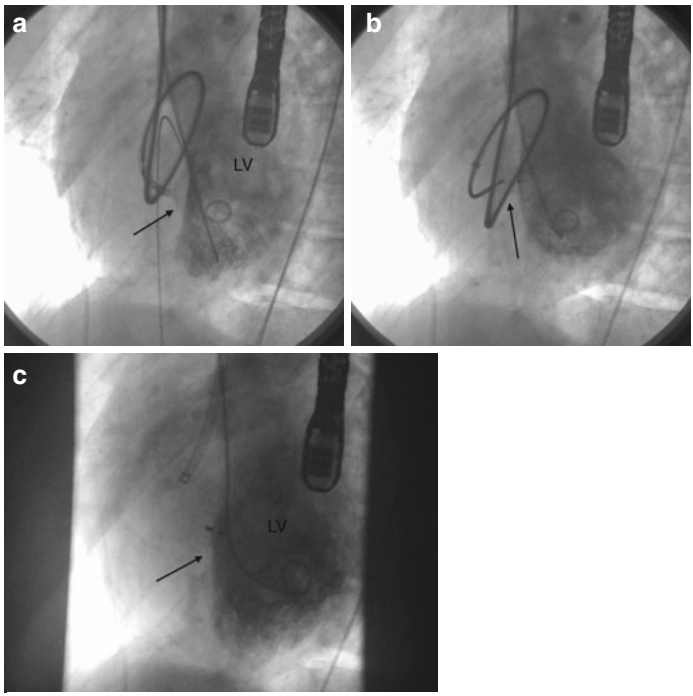

Fig. 28.5 Left ventricular angiogram: (**a**) The system is withdrawn towards the septum (*arrow*); (**b**) the central waist and the proximal disc are deployed (*arrow*); (**c**) a test angiogram is done to verify the correct position of the device (*arrow*). *LV* left ventricle

Echocardiographic views are also very important to confirm the position of the two discs on left and right side of the septum, respectively, and the central waist within the muscular septum.

The device is then released.

A final angiogram is performed approximately 10–15 min afterwards to assess the position of the device and the possible residual shunt (Fig. 28.5c)

Patients receive acetyl salicylic acid (3–5 mg/kg/daily maximum 300 mg/daily) for 6 months and are asked to follow strictly endocarditis prophylaxis.

A similar approach may be used to close multiple muscular VSDs.

28.6.1 Alternative Techniques

28.6.1.1 Retrograde Approach

This approach can be used in adults and older children in whom a 7–8-French arterial introducer can be used safely.

The VSD is crossed from the left ventricle with the help of a soft 0.035″ J Tip Terumo 260-cm exchange wire introduced through a 5-Fr Judkins right coronary artery catheter.

The wire is then advanced in the pulmonary artery. The catheter is exchanged with a 80-cm delivery sheath (AGA medical) over the wire to the right ventricle apex. The wire and dilator are removed slowly in order to avoid air suctioning.

The chosen device is prepared and advanced into the long sheath. The distal disc is opened in the RV apex paying attention to the ventricular wall and tricuspid wall.

The whole system is then pulled back to approximate the interventricular septum. The sheath is further withdrawn to open the proximal disc onto the left ventricular surface of the interventricular septum.

Left ventricular angiograms and echocardiographic evaluations are performed to confirm the position of the device and the absence of complications.

The device is unscrewed from the delivery cable, and angiograms are performed in the ascending aorta and left ventricle to confirm the final position of the device to search for residual shunt and to check aortic valve function.

28.6.1.2 Hybrid Approach

A hybrid approach has been developed to overcome the risks of the two procedures (percutaneous closure may be hazardous due to vascular access and haemodynamic tolerance of the procedure, and a surgical approach needs extracorporeal circulation and may be associated to significant morbidity and mortality in particular in case of apical defects) in smaller infants (less than 6 kg).

The chest and the pericardium are opened, under TEE control; an 18-gauge needle is used to puncture the right ventricle free wall.

A 5-0 polypropylene purse string suture is placed around the puncture site.

The needle is introduced into the right ventricular cavity pointing towards the VSD.

A 0.025 short guide wire is passed through the needle and the VSD in the left ventricle.

Over the wire, a short sheath is advanced to the left ventricle cavity.

A proper-sized Amplatzer muscular VSD device is delivered using TEE monitoring

28.7 Catheterization and Haemodynamic Evaluation for PMVSD Closure

Angiographies are performed using 60° left atrial oblique plus 20° cranial view (Fig. 28.6).

An angiogram of the ascending aorta is also performed in 50° left atrial oblique view to check for aortic insufficiency.

The size of the defect, and its relationship to the aorta, is confirmed by 2–3D TEE (Fig. 28.7a, b).

The defect is crossed from the left ventricle by using a right Judkins or a right Amplatzer catheter and a Terumo wire. The catheter is advanced to the pulmonary arteries or to the SVC/IVC;

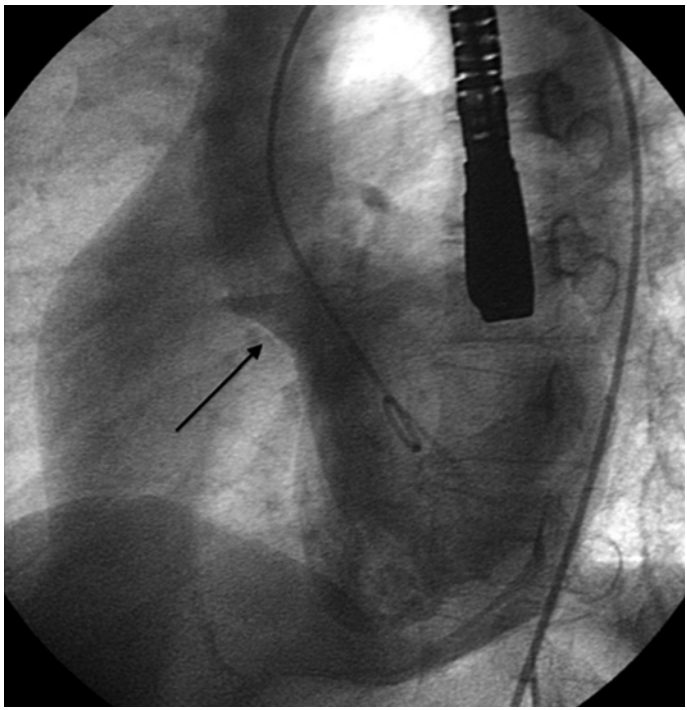

Fig. 28.6 Left ventricular angiogram: the *black arrow* shows the PMVSD

the Terumo wire is then replaced by the soft exchange noodle wire (a dedicated 300-cm exchange guide wire, AGA Medical Corporation, Golden Valley, MN), snared with a GooseNeck Snare, and exteriorized from the femoral vein (arteriovenous circuit) (Fig. 28.8a, b).

The AGA braided sheath is advanced over the wire up to the ascending aorta.

Use a "kissing" technique if some resistance are encountered: both the tip of the sheath and of the arterial catheter over the wire must be in contact and pushed-pulled together.

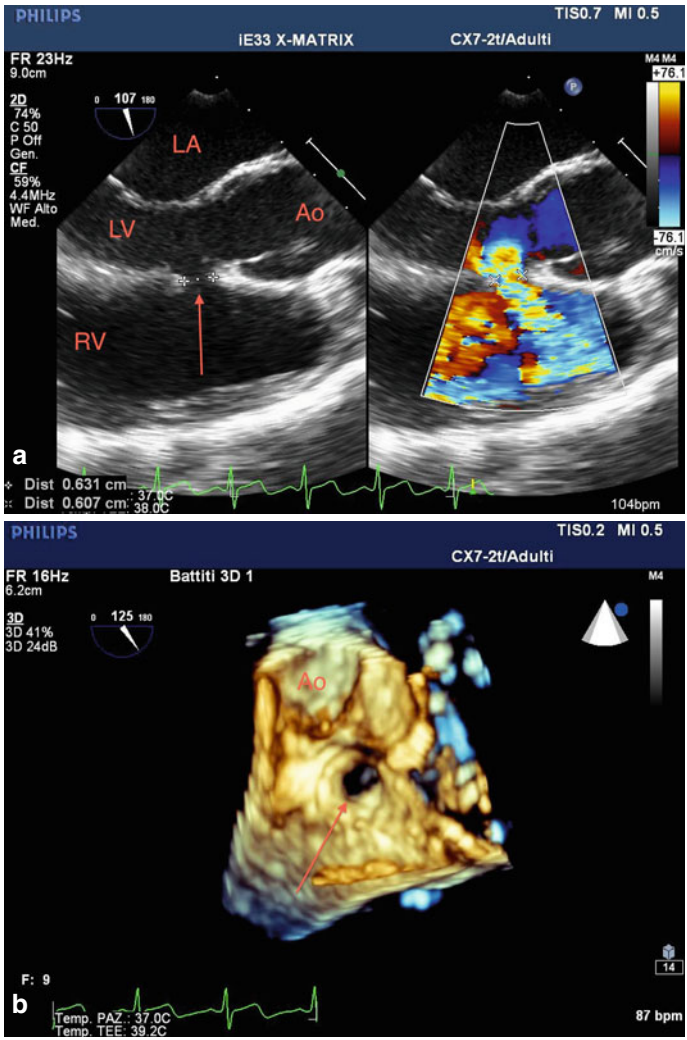

Fig. 28.7 (**a**) 2D-TEE long-axis view: *arrow*, PMVSD; *RV* right ventricle, *LV* left ventricle, *LA* left atrium, *Ao* aorta. (**b**) 3D-TEE: *arrow*, PMVSD; *Ao* aorta

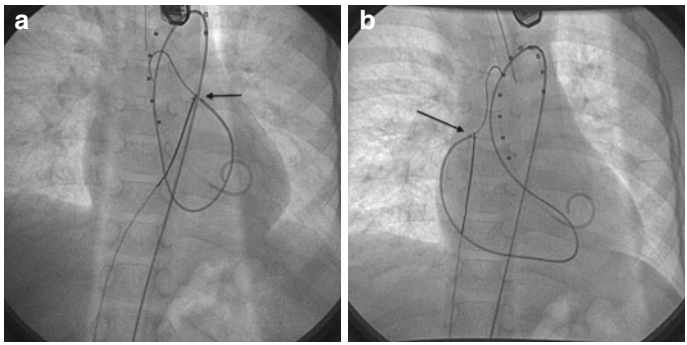

Fig. 28.8 The wire is snared with a GooseNeck Snare in the left pulmonary artery (**a**) or in the superior caval vein (*arrow*) (**b**) and exteriorized from the femoral vein (*arrow*) (arteriovenous circuit)

When the long sheath is in ascending aorta, hold the guide wire circuit, withdraw the dilator of approximately 10 cm, withdraw slowly the sheath and advance the arterial catheter; make a loop of the wire and push it into the left ventricular apex.

The sheath is then advanced over the wire until it reaches the apex of the left ventricle, and the wire is gently removed (Fig. 28.9).

The device, having been sized at equal to or 1 mm larger than the size of the defect, is secured on the delivery cable, and the flat part of the microscrew is aligned with the flat part of the capsule of the pusher catheter.

The device is advanced up to the tip of the sheath, and the entire system is withdrawn to the left ventricular outflow tract (Fig. 28.10).

When the left disc is deployed, echocardiographic monitoring is of paramount importance to confirm normal function of both mitral and aortic valve.

The platinum marker of the distal disc should point downwards.

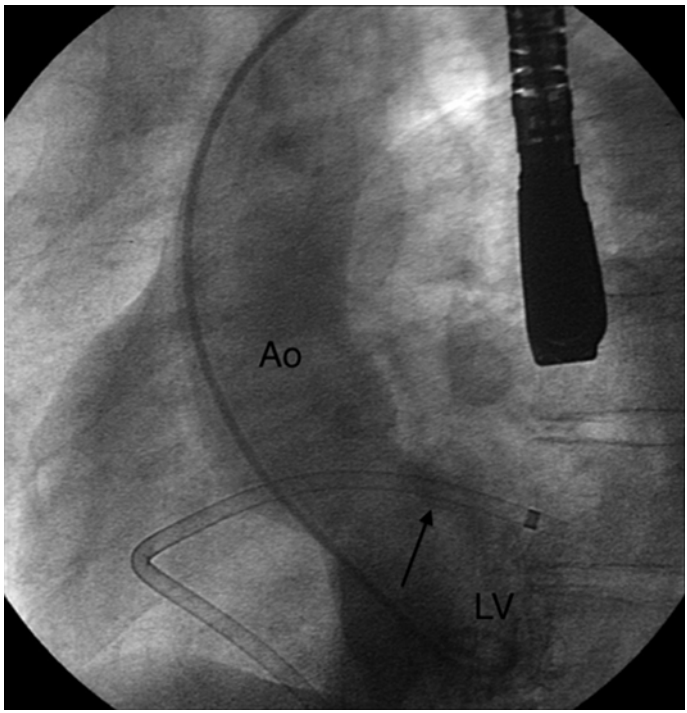

Fig. 28.9 The long sheath (*arrow*) into the left ventricle (*LV*). *Ao* aorta

The proximal disc is then deployed on the right side of the septum, and angiographic testing is done before releasing the device (Fig. 28.11a–c).

When it is difficult to achieve the position of the braided sheath towards the left ventricular apex, the sheath can be left in the ascending aorta and the left ventricular disc opened under the aortic valve while coming with the sheath from the aorta. Then the right ventricular disc is opened by advancing the delivery cable (Fig. 28.12a, b).

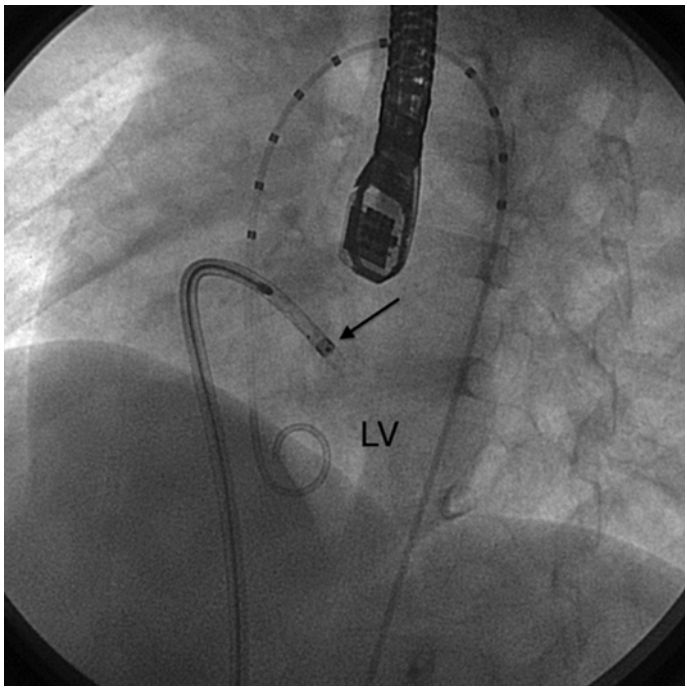

Fig. 28.10 The device is up to the tip (*arrow*) of the long sheath, and the entire system is withdrawn to the left ventricular outflow tract. *LV* left ventricle

After 10–15 min, the left ventricular angiogram and aortogram are repeated to assess possible residual shunting or aortic regurgitation (Fig. 28.13a, b). Throughout the procedure, the electrocardiogram is carefully screened in order to assess the occurrence of abnormalities of atrioventricular conduction or tachyarrhythmias.

The most common morphological variation is the presence of an aneurysm of the ventricular septum (Fig. 28.14).

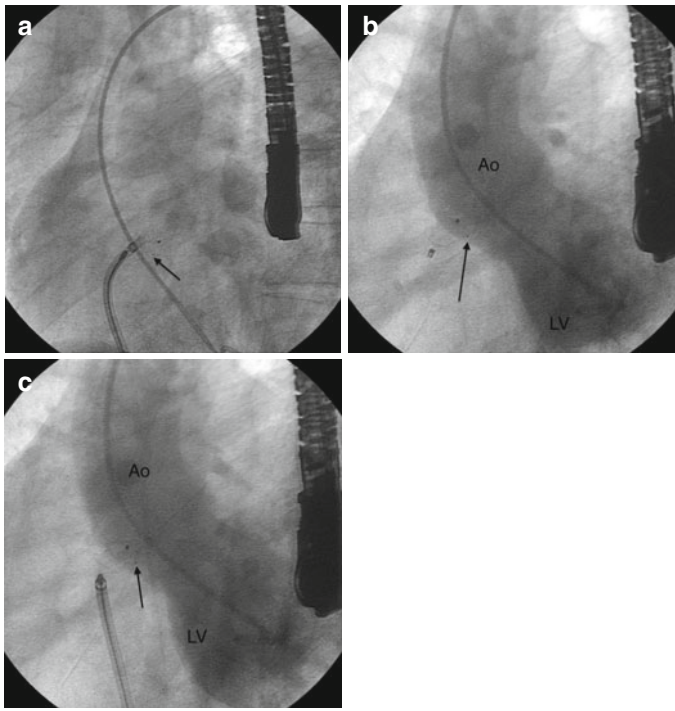

Fig. 28.11 Left ventricular angiogram: (**a**) the left disc is open and withdrawn to the interventricular septum. (**b**) The proximal disc is then deployed on the right side of the septum, and angiographic testing is done before releasing the device. The *arrow* shows the platinum marker of the distal disc pointing downwards. (**c**) The final angiography shows the complete closure of the defect. *Ao* aorta, *LV* left ventricle

Better try to close the true anatomical hole with the more appropriate device (muscular for perimembranous AGA device).

If the redundant tissue of the aneurysm is relatively small, the device could cover the hole along with the aneurysm (Fig. 28.15).

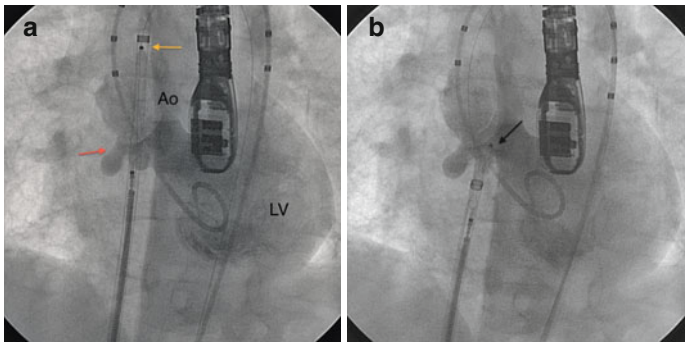

Fig. 28.12 Left ventricular angiogram: (**a**) the sheath can be left in the ascending aorta (*yellow arrow*). The *red arrow* shows the aneurysm of the PMVSD. (**b**) The left ventricular disc opened under the aortic valve while coming with the sheath from the aorta (*arrow*). *Ao* aorta, *LV* left ventricle

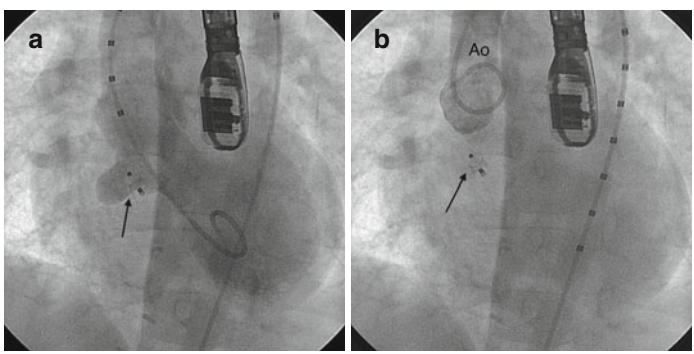

Fig. 28.13 The left ventricular angiogram (**a**) and aortogram (**b**) are repeated at the end of the procedure to assess possible residual shunting or aortic regurgitation. The *arrows* show the device setting into the aneurism. *Ao* aorta

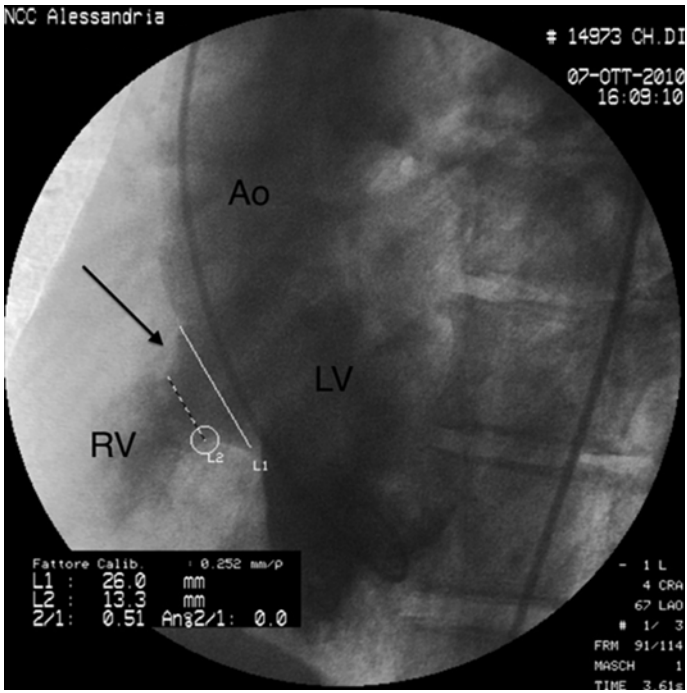

Fig. 28.14 Left ventricular angiogram showing a PMVSD with an aneurysm (*arrow*). *Ao* aorta, *LV* left ventricle, *RV* right ventricle

In case of very large aneurysms, the device may be implanted within the aneurysm itself, with the aim of closing the true anatomical hole and not to place the device at the "entrance" on the left ventricular side, avoiding insertion of a dangerously oversized device.

In case of conic shape of the aneurism, different devices (PDA I AGA Medical Corporation, St. Jude, MN, or a Nit-Occlud® Lê VSD pfm medical ag.Köln, Germany) may be taken into consideration.

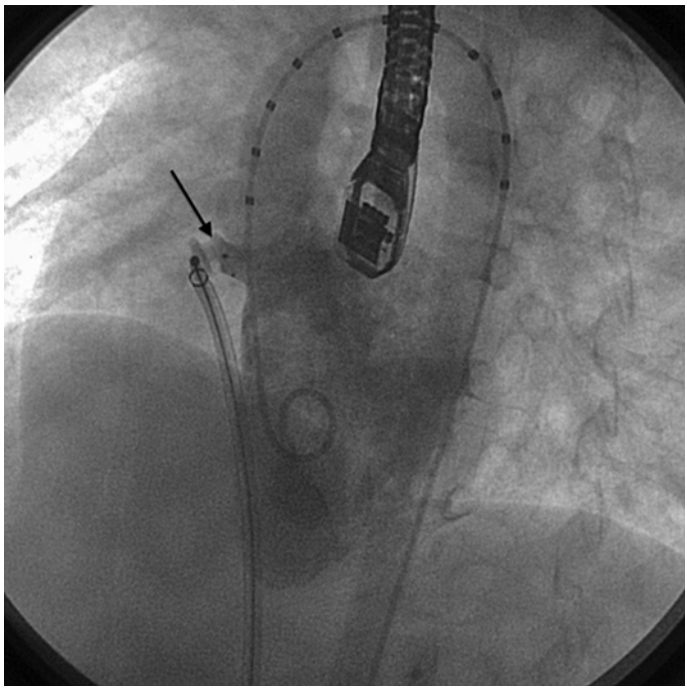

Fig. 28.15 Left ventricular angiogram. The *black arrow* shows the device deployed into the aneurysm

28.8 Specific Technical Aspects for Postsurgical Residual VSD

Balloon sizing of the defect: due to the varied anatomy of the substrate (presence of patches and patch leaks), sizing at the TEE and angiography can be more difficult. Balloon occlusion of the shunt and assessment with TEE and angiography provide significantly better understanding of the shunt size and site.

Aortic retrograde approach: the majority of these VSDs are located in the muscular septum, with a potential risk of the sheath passing through or under a trabeculation of the RV.

The standard anterograde approach may be more difficult (presence of the surgical patch, less space in the subaortic region to deploy the LV disc and increased risk of complications).

A retrograde approach may overcome these issues.

28.9 Complications

Complications in closure are reported in 1.3 up to 5 %.

Major procedure related complications include:

- Embolization of the device (likely to be related to the learning curve or lack of experience by the operator. An underestimated size device usually was implanted). The device can be retrieved, and a second device can be implanted.
- Cardiac perforation (be careful placing and moving guide wires, delivery sheath and the device).
- Stroke (frequently related to air embolism).
- Deaths (rare).
- Haemolysis, frequently transient.
- Aortic regurgitation (related to PMVSD closure).
- Disturbances of conduction (related to PMVSD closure). Complete heart block (CAVB) is a serious complication in children (not in adult patients). It may occur acutely (transiently, during the procedure or permanent) or months after the procedure but permanently. Implantation of a pacemaker may be required.
- The exact mechanism of CAVB remains unclear (inflammatory reaction, formation of scarring in the conduction system, impingement against the vascular conduction system supply). To reduce the risk of CAVB, avoid oversizing the device size by more than 1 mm.

Suggested Reading

1. Chessa M, Carminati M, Cao QL, Butera G, Giusti S, Bini RM, Hijazi ZM (2002) Transcatheter closure of congenital and acquired muscular ventricular septal defects using the Amplatzer device. J Invasive Cardiol 14:322–327
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