Chapter 66
Transcatheter Perimembranous VSD Device Closure

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Abstract The prevalence of VSD in adults is lower than the other defects with left to right shunts. When is detected in adulthood, it may cause LV volume overload or can be lead to Eisenmenger syndrome. It is off-label to close the perimembranous type (the most common type) percutaneously. But by recent advances in device types, it seems that trans-catheter closure of this type of VSD can be done safely with low complications.

History

The patient was a 32 years old man who was referred with complaints of exertional dyspnea and orthopnea since 2 years ago and recently with nocturnal palpitation. In TTE a large perimembranous VSD with mild AI was detected.

Isolated VSD accounts for 37% of all congenital heart disease in children. Because as many as 90% eventually closed spontaneously; the incidence is significantly lower in adults. Multiple synonyms have been classified and categorized VSDs into four major groups:

- Type 1: (Infundibular, outlet) This VSD is located below the semilunar valves (aortic and pulmonary) in the outlet septum of the right ventricle above the crista supraventricularis that is why sometimes also referred to as supra-cristal. It is the most uncommon type representing only 6% of all VSDs with the exception being in the Asian population where it accounts for approximately 30%. Aortic valve prolapse and regurgitation are common because of loss of support of the right and/or the noncoronary cusps of the aortic valve. It is unusual for these defects to close spontaneously.
- Type 2: (membranous) This VSD is, by far the most common type, accounting for 80% of all defects. It is located in the membranous septum inferior to the

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crista supraventricularis. It often involves the muscular septum when it is commonly known as perimembranous. The septal leaflet of the tricuspid valve sometimes forms a “pouch” that reduces the shunt and can result in spontaneous closure.
- Type 3: (Inlet or atrio-ventricular canal) This VSD is located just inferior to the inlet valves (tricuspid and mitral) within the inlet part of the right ventricular septum. It only represents 8% of all defects.
- Type 4: (muscular, trabecular) This VSD is located in the muscular septum, bordered by muscle usually in the apical, central, and outlet parts of the interventricular septum. They can be multiple, assuming a “Swiss cheese” appearance. They represent up to 20% of VSDs in infants. However, the incidence is lower in adults due to the tendency of spontaneous closure [1].

The presentation of unrepaired VSDs is dependent on the presence of hemodynamically significant shunt; that it is directly related to the size of the defect. Small VSDs only lead to minimal left-to-right shunt without hemodynamic effects; they are usually asymptomatic. Medium size VSDs result in a moderate LV volume overload and mild PAH; they present late in childhood with some congestive symptoms. Those with large defects develop CHF early in childhood due to severe LV overload and severe PAH and finally Eisenmenger syndrome which occurs in approximately 10–15% of patients. These patients present with dyspnea, cyanosis, syncope, palpitation, and sudden cardiac death.

Diagnostic Work-Up

In small size VSDs, a loud and harsh pansystolic murmur is heard in LSB, but it gets softer with increasing the size of the defects, and eventually, the typical murmur will be absent in the large ones with accentuated P2 sound. ECG shows LVH in moderate size defects. In those with Eisenmenger syndrome, right axis deviation and RVH is seen. TTE is the essential tool for the detection of VSDs (defect size, numbers, location, the relationship of the VSD with other structures, hemodynamic effects, LV and RV size and function, associated anomalies, and PAP).

Indications and Contraindications of VSD Closure (Table 66.1)

RHC

- Qp/Qs: 2.5
- PVR: 2 WU
- SVR: 120 WU
- PAP: 55/20 mm Hg

Table 66.1 AHA/ACC 2018 guideline

COR	LOE	Recommendations
Therapeutic		
I	B-NR	1. Adults with a VSD and evidence of left ventricular volume overload and hemodynamically significant shunts (Qp:Qs ≥ 1.5:1) should undergo VSD closure, if PA systolic pressure is less than 50% systemic and pulmonary vascular resistance is less than one third systemic.[54.1.3-1]
IIa	C-LD	2. Surgical closure of perimembranous or supracristal VSD is reasonable in adults when there is worsening aortic regurgitation (AR) caused by VSD.[54.1.3-1.54.1.3-2]
IIb	C-LD	3. Surgical closure of VSD may be reasonable in adults with a history of IE caused by VSD if not otherwise contraindicated.[54.1.3-3]
IIb	C-LD	4. Closure of a VSD may be considered in the presence of a net left-to-right shunt (Qp:Qs ≥ 1.5:1) when PA systolic pressure is 50% or more than systemic and/or pulmonary vascular resistance is greater than one third systemic.[54.1.3-4-54.1.3.6]
III: Harm	C-LD	5. VSD closure should not be performed in adults with severe PAH with PA systolic pressure greater than two third systemic, pulmonary vascular resistance greater than two thirds systemic and/or a net right-to-left shunt.[54.1.3-7-54.1.3.9]

- RVSP: 55/12 mm Hg
- mRAP: 12 mm Hg
- So2: 95%

Transcatheter PM-VSD Device Closure

Right femoral artery and vein sheaths (6F) were inserted then IV Heparin was completed to achieve ACT > 200 s. After LV injection in LAO 60-Cranial20 in order to profile the defect (Fig. 66.1a), Under general anesthesia, in retrograde approach, the VSD was crossed from LV via JR and 0.035 inch hydrophilic guide-wire (in some cases based on the location of the defects, another tip angled catheters like IMA or cutoff pigtail are used). After crossing the defect with the wire and the catheter that was being passed through it and inserted in the LPA, the wire was exchanged with long 260 cm Terumo wire. Through venous access,

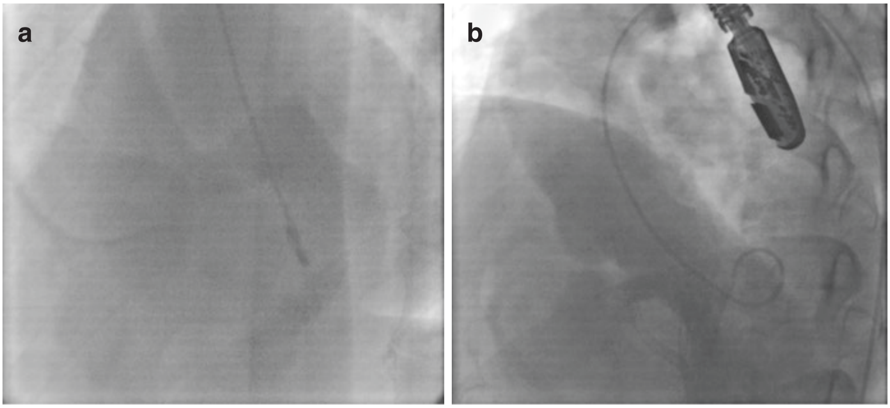

Fig. 66.1 (**a**, **b**) LV and aortic root angiography showed, a large perimembranous VSD with RV opacification

a pigtail catheter without guide-wire was inserted in the same pulmonary artery that the Terumo wire was passed (to reduce the interference with the Tricuspid valve chordae), then the pigtail was exchanged with JR guiding catheter. To make an arterio-venous loop, the guide-wire was then snared and exteriorized via venous access. A Delivery sheath (10F) was advanced carefully to RV, LV, and aortic arch with Kissing catheter technique (Fig. 66.1b). Then dilator was removed from the vein access and guide-wire and the end hole catheter were removed from the arterial line. Under TEE guiding and expert echo-man, according to the defect size (8 mm - largest diastolic phase on LV side) and acceptable distance between the defect and the aortic cusps, Membranous VSD Occluder (Cera Lifetech symmetric) device 10 mm was chosen. After the device was screwed to the delivery cable and de-aired, it passed through the long sheath and under TEE and fluoroscopy guidance, first Left disk was partially opened in ascending aorta and gently pulled back through the AV to the LV (Fig. 66.2a) and slowly, by retracting the delivery sheath and fully deploying distal disk at the LV side of VSD, the entire system was then pulled back into the defect (Fig. 66.2b) and the sheath was retracted to deploy the waist and finally after confirming the right position of the device by TEE and LV injection, the proximal (Right disk) was deployed (Fig. 66.2c). The operator must fully check the position and stability of the device without any compromising of the surrounding structures. If there is any doubt about the impingent of the valves or other structures, the device should be recaptured. If the device position is correctly confirmed by the TEE and angiography, it can be released by unscrewing it using pin vice (Fig. 66.3a, b). In this case, the AI severity or TR was not increased without any conduction abnormality.

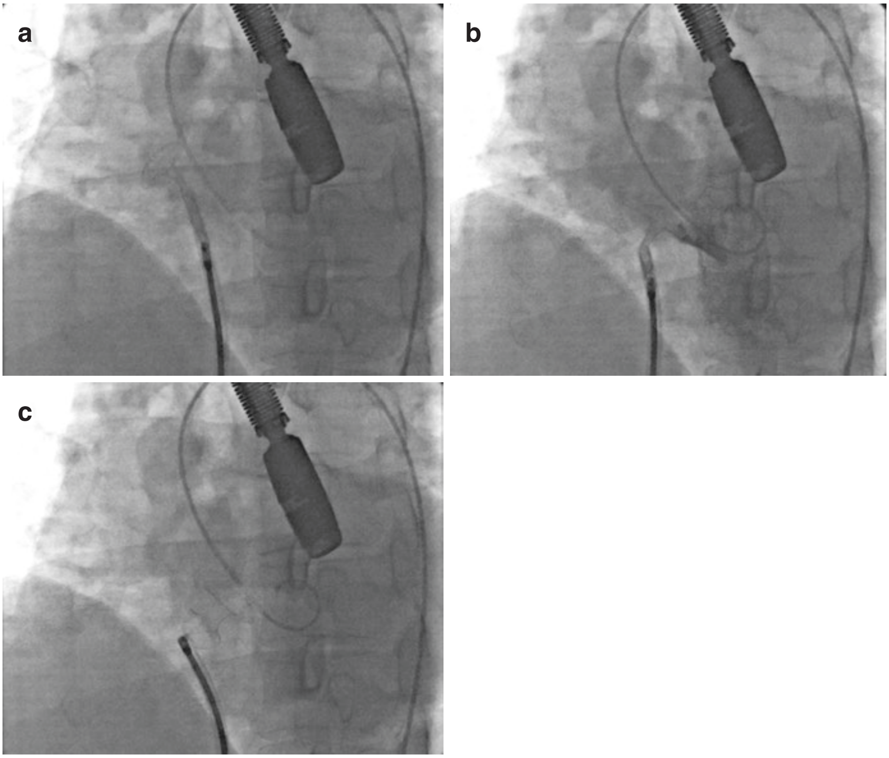

Fig. 66.2 (**a**, **b**, **c**) According to the defect size (8 mm - largest diastolic phase on LV side) and acceptable distance between the defect and the aortic cusps, Membranous VSD Occluder (Cera Lifetech symmetric) device 10 mm was chosen. first Left Disk was partially opened in the ascending aorta and gently pulled back through the AV to the LV and slowly, by retracting the delivery sheath and fully deploying distal disk at the LV side of VSD, the entire system was then pulled back into the defect and the sheath was retracted to deploy the waist and finally after confirming the right position of the device by TEE and LV injection, the proximal (Right Disk) was deployed

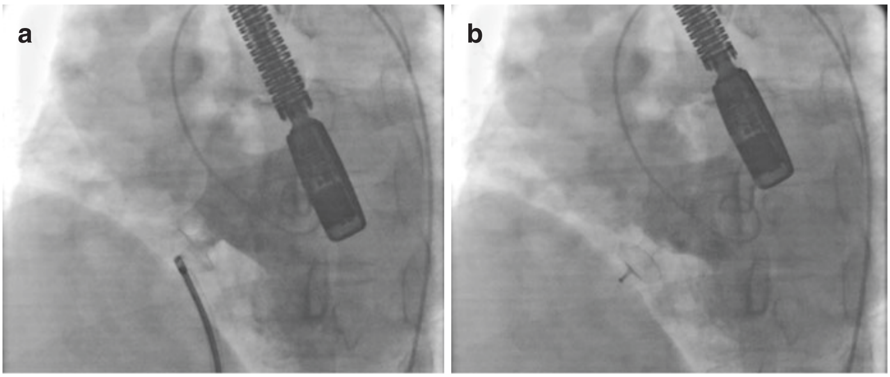

Fig. 66.3 (**a**, **b**) After confirming the right position of the device with no interaction with surrounding structures, the device was released and final LV angiography showed tiny residual shunt

Conclusion

Percutaneous closure of PM-VSD is a challenging procedure, owing to variable anatomical morphology, proximity to valves and conduction systems as well as complex manipulation process. The major key to improve the results of this treatment, while minimizing complications (such as: CHB, device embolization, valvular malfunction, and hemolysis) consists in the careful case and device selections as well as accurate defect sizing strategy [2].

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