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Transcatheter Closure of Congenital VSDs: Tips and Tricks

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Additional information is available at the end of the chapter

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Abstract

Nowadays transcatheter device closure of ventricular septal defects (VSDs) is an attractive and feasible alternative to surgical closure of congenital VSDs. Isolated congenital VSDs constitute the most common form of congenital heart disease (CHD) in infants and children and account for 20–30% of all types of cardiac malformations. Most of the VSDs are located in the membranous portion of the ventricular septum (perimembranous VSDs). There are also less common types of VSDs located in the muscular portion (muscular VSDs), below the pulmonary valve (subpulmonary or supracristal VSDs), and near the junction of the tricuspid and mitral valves (inlet type VSDs). Indications for closure of VSDs include a hemodynamically significant left to right shunt and prevention of long-term complications, including pulmonary hypertension, progressive ventricular dilatation, aortic insufficiency, double-chambered right ventricle, and endocarditis. In this chapter, we review the technical details for achieving a successful procedure, as well as some tips and tricks on using off-label devices during transcatheter approach in VSD closure.

Keywords: congenital VSD, transcatheter device closure

1. Introduction

Ventricular septal defect (VSD) is the most common congenital heart defect and accounts for approximately 20% of all forms of congenital heart disease as an isolated lesion with incidence increasing up to 40% in case of multiple congenital heart defects [1]. Perimembranous VSDs are the most common form (70%), and muscular (15–20%) and sub arterial (5%) are less common (**Figure 1**). The size of the defect determines the size of the left to right shunt, which affects the hemodynamic state from negligible to cardiac failure and mild to severe

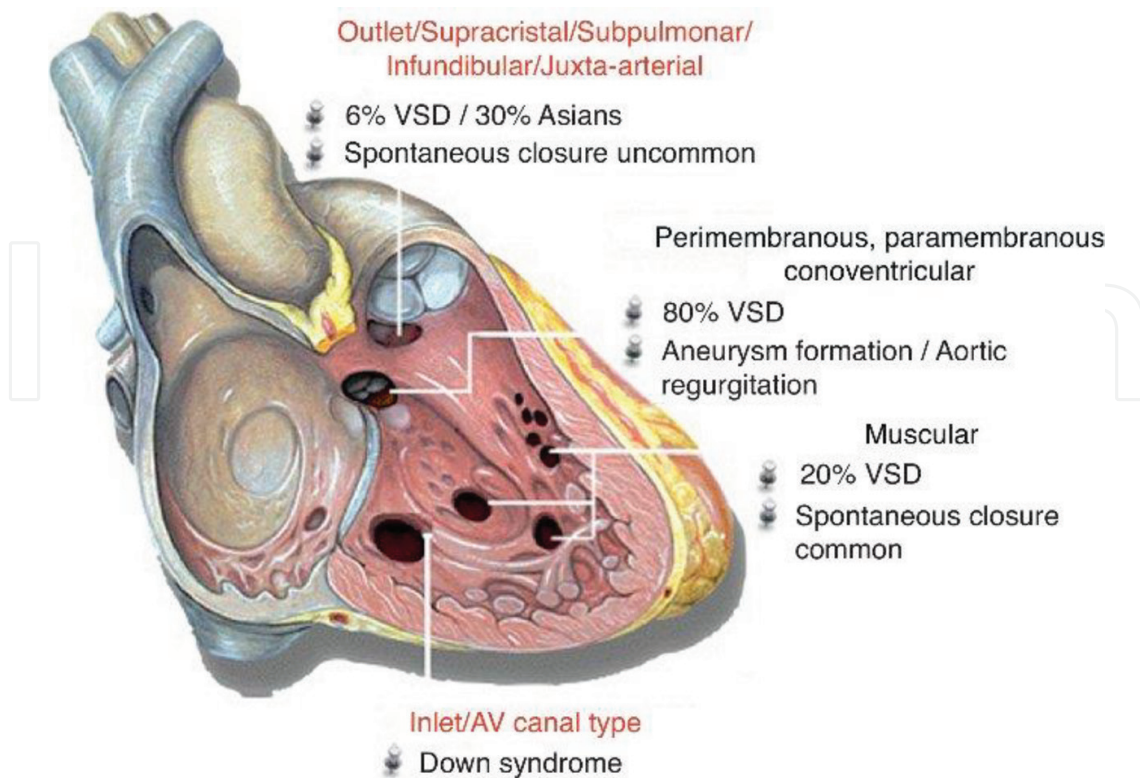


Figure 1. Prevalence of different forms of VSDs (picture courtesy: Patrick J Lynch; C. Carl Jaffe Yale University).

pulmonary hypertension. Although most of the larger defects persist through adulthood, some smaller defects have a high likelihood of spontaneous closure. There are some long-term complications of VSDs including prolapse of aortic cusps with regurgitation, infective endocarditis, arrhythmias, and pulmonary hypertension, which may lead to pulmonary vascular obstructive disease or Eisenmenger syndrome.

Traditionally, closure of ventricular septal defects (VSDs) has been a surgical procedure for over 50 years with a low operative mortality and postoperative morbidity. However, in 1988, Lock et al. [2] reported the results of transcatheter VSD closure using the Rashkind double umbrella device in six patients with congenital and acquired VSDs. Transcatheter closure of VSD as an alternative to surgery has now gained increasing acceptance due to a comparable success rate and low risk of complications. This approach has several advantages, such as avoidance of sternotomy and cardiopulmonary bypass, with less pain and no scar and shorter hospital stay, as well. There are also some disadvantages like the need for X-ray and contrast media injections.

Although surgical treatment remains the standard approach for VSDs, percutaneous device closure has brought hope to be a safe and effective treatment with a high rate of success.

2. Historical aspects

Transcatheter closure of ventricular septal defects was first described by Rashkind when he used a single-disc device to perform this in dogs.

Lock et al. used Rashkind double disc PDA umbrella in human subsequently. Six of the seven devices were implanted successfully in his series, while the seventh embolized into the pulmonary artery. Goldstein used clamshell occluder to close the VSDs. Gianturco coils, Amplatzer membranous and muscular devices, buttoned device, wireless devices (detachable steel coils, detachable balloon, and transcatheter patch), cardioSEAL/STARFlex devices, Nit-occlud (Nickel-Titanium Spiral Coil), and Amplatzer Duct Occluder I and Amplatzer Duct Occluder II devices were used for transcatheter occlusion of VSDs subsequently.

Postmyocardial infarction VSDs were also closed percutaneously using Rashkind double disc PDA umbrella, clamshell, CardioSEAL or STARFlex, Amplatzer septal occluder, Amplatzer duct occluder, and Amplatzer post-infarct muscular VSD (PIMVSD) successfully.

Amplatzer muscular VSD and PIMVSD occluders, Qwik-Load and STARFlex Septal Occlusion System, and CardioSEAL Septal Occlusion System are now approved by FDA, but there are multiple devices that have been used to close VSDs so far, which include VSD Le Coils, Muscular and Membranous VSD devices of Occlutech Co., and many Chinese symmetrical and Asymmetrical VSD occluders and the Amplatzer Duct occluder with Duct Occluder II (St. Jude Medical, Inc.) as well. The use of the Chinese devices is most commonly reported from China itself. Although the Amplatzer membranous VSD occluder was found useful, development of heart block precipitated its removal from clinical trials in the USA. Other devices are in clinical trials in either the USA or abroad [1].

3. Indications

Different patients with VSDs may present with different scenarios. Most muscular VSDs and a few perimembranous type may close spontaneously in early month of life. Some large unrestrictive VSDs may cause heart failure and failure to thrive in small babies. These infants need a large device for interventional closure, which makes the procedure difficult and unsafe. So this group of patient often refers to surgeons.

Although closure of VSDs in patients with aortic valve prolapse or AI remains controversial and most of this patients with trivial AI are now referred to surgeons, Le VSD Coil (pfm medical ag, Koln, Germany) occluder has recently used in such condition with good results [2].

VSDs are considered eligible for transcatheter device closure in the presence of one or more of the following indications:

- Evidence of heart failure not controlled by medical therapy
- Pulmonary to systemic blood flow ratio greater than 1.5 ($Q_p/Q_s > 1.5$)
- Evidence of left heart volume over load
- History of previous endocarditis

3.1. Contraindications

- body weight less than 6 kg
- pulmonary vascular resistance index greater than 7 WU/m^2 , unresponsive to oxygen

- PMVSD extending to the inlet
- the presence of additional lesions requiring surgical intervention (ToF)
- when parents prefer surgical intervention.

Pulmonary hypertension is defined as a mean pulmonary artery pressure of 20 mm Hg or greater based on the original natural history studies [3].

There are some other considerations like distance from the edge of the VSD to the semilunar valves:

- <2 mm—for the Amplatzer membranous VSD occlude
- <4 mm—for the Nit-Occlud VSD

4. Preprocedure assessment

Echocardiography can provide valuable information on the number, the location, the size, and the relationship of the VSD to the adjacent structures. In addition, transesophageal and 3D-echocardiography are now widely available and may provide additional information of unusual VSDs.

5. Technique

5.1. General considerations

Compared to patent ductus and atrial septal defect closure, VSD closure is considerably more complicated. It is thus important to recognize that this procedure should only be undertaken in well-equipped units with sufficient skill, knowledge, and surgical backup.

- Anesthesia: the procedure may be performed under general anesthesia, although sedation can be used.
- Imaging: most of the centers use continuous transesophageal echocardiography (TEE), but skilled echo cardiographers may use transthoracic echo during the procedure.
- Catheterization: a comprehensive evaluation should be done during angiography to obtain different views at multiple angles of the VSD and a complete study for valvular function and regurgitation as well.
- Access: although in some certain muscular VSDs, the right internal jugular vein access may be used, and femoral artery and vein are usually gained as the main access. Alternatively, in small patients who making an arteriovenous circuit may cause hemodynamic instability when long stiff sheath is placed across the tricuspid valve, a hybrid approach can be used.

In some selected older children with some difficulties, the VSD may be approached retrogradely from the femoral artery. Activated clotting time is maintained above 200 seconds throughout the procedure.

- Device selection: the device is usually selected 1–2 mm larger than the maximal diameter of the defect as assessed by TEE and angiography. Balloon sizing is hardly ever used, since the inter-ventricular septum is regarded to be a nonstretchable structure. There are some other important considerations in device selection but paying attention to the location, shape, morphology, and length and thickness of the edges of defect are of the most important factors (Figures 2 and 3).
- Crossing the VSD: normally, the VSD is crossed from the left ventricle using a Judkins right coronary artery catheter, but a variety of tip angled catheters can be used based on the defect location and shape like Bern, Cobra or a cutoff Pigtail. An exchange guidewire like Terumo or Noodle wire is then placed into the left or right pulmonary artery via the VSD crossed catheter (Figure 4B).

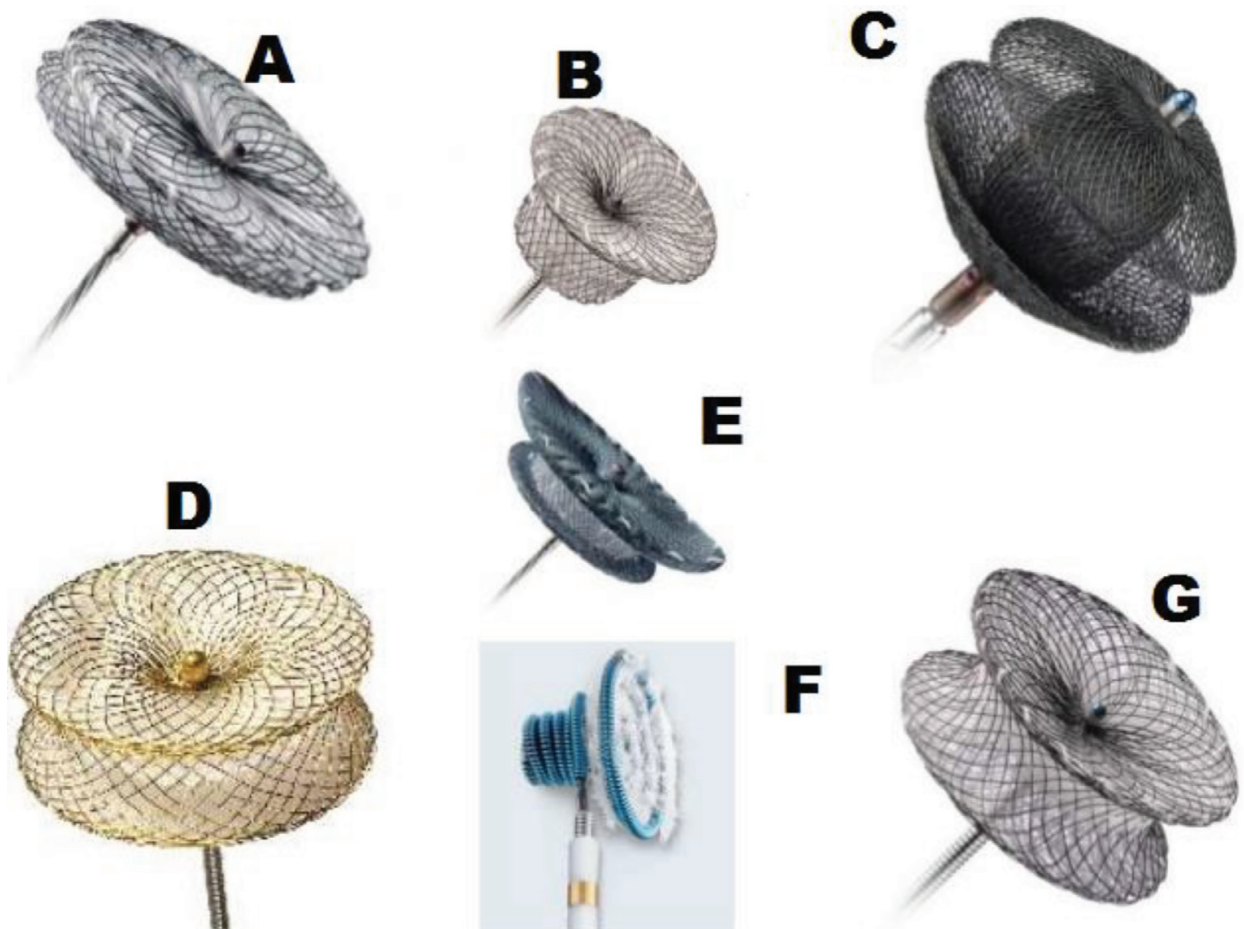


Figure 2. (A) Amplatzer membranous VSD device, (B and C) Amplatzer Duct Occluder and Amplatzer Duct Occluder II devices. (D and G) Amplatzer and Occlutech muscular VSD device. (F) Nit-Occlud (Nickel-Titanium Spiral Coil) and (E) asymmetric membranous VSD device were used for transcatheter occlusion of VSDs.

- Arteriovenous guide wire loop: to make an arteriovenous wire loop, the guide wire is then snared and pulled out via the right femoral vein. A delivery sheath will be then advanced via the femoral vein access into right ventricle into the aorta carefully. Undue tension on the arteriovenous loop and VSD may cause rhythm disturbances. To avoid direct contact of the guide wire with the VSD when crossed by the sheath, a “kissing catheter technique” should be used. The delivery sheath is then positioned to a suitable position in the left ventricle or descending aorta (**Figure 4C,D**).
- The device: the proper size and suitable shape device is then screwed to the tip of the delivery cable and advanced to the cavity of left ventricle or descending aorta via the long sheet with special care to avoid the occurrence air embolism. The LV disc is first deployed within the LV chamber or descending aorta and gently pulled back to the intra-ventricular VSD under echo or angiographic guidance. The waist of the device and right ventricular disc are then deployed, respectively. Careful attention should be paid to good positioning and stability of the device and any potential compromising of the adjusting structures. In the case of any impingement of valves or other structures, the device can still be recaptured into the sheath and repositioned gently. Once proper positioning has been achieved, the device may be released by unscrewing it counter clockwise using the pin vice. After release, confirmation of correct positioning should be established using TEE or TTE and angiography (**Figures 4F-I** and 5).

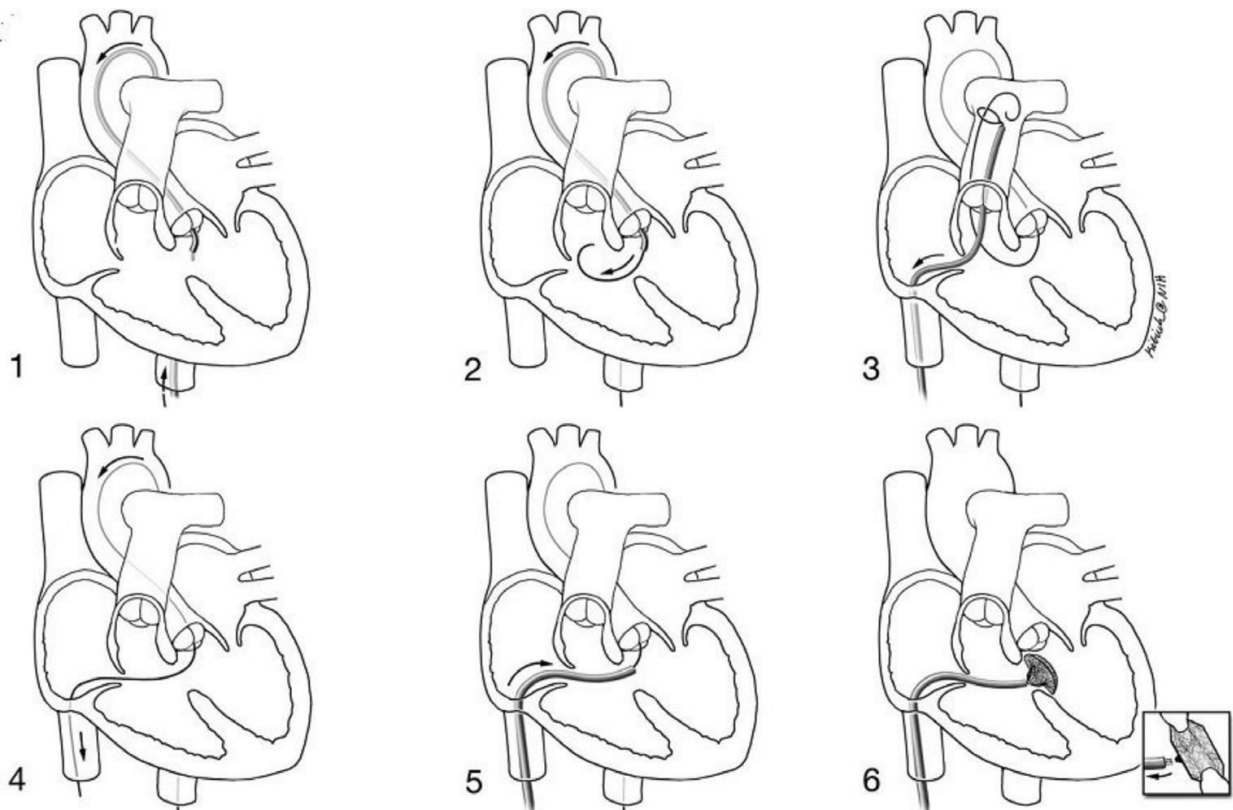


Figure 3. Schematic depictions of conventional technique. 1—Transaortic access into LV chamber retrogradely. 2—Crossing a guide wire from LV to RV chamber retrogradely. 3—Snaring the trans-aortic guide wire from the pulmonary artery. 4—Making an arteriovenous loop to provide a way to deliver the VSD delivery sheath. 5—The delivery sheath is advanced antegradely across the VSD. 6—The device is fixed in position and released. (Courtesy of Lydia Kibiuk, NIH Medical Arts and Photography Branch).

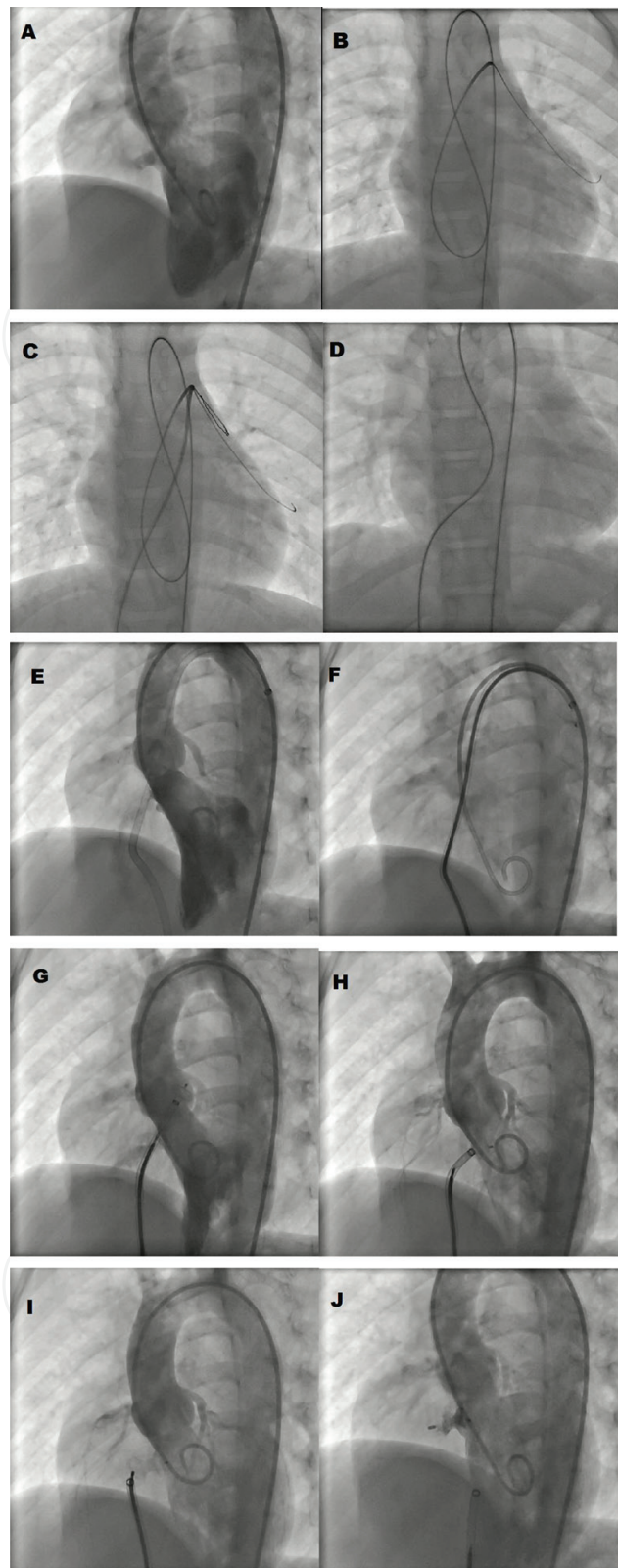


Figure 4. Stepwise percutaneous VSD closure technique using CaridoFix Muscular VSD Occluder. (A) Small upper muscular VSD. (B) Crossing the VSD and putting the wire in pulmonary artery. (C) Snaring the pulmonary wire via venous access. (D) Making the AV loop from arterial and venous access. (E) Crossing the VSD by long sheet and into the descending aorta. (F) Releasing the first disc of the device. (G) Pulling back the device. (H) Fixing the device to the septum. (I) Releasing the second disc of the device. (J) Complete deploying and releasing of the device.

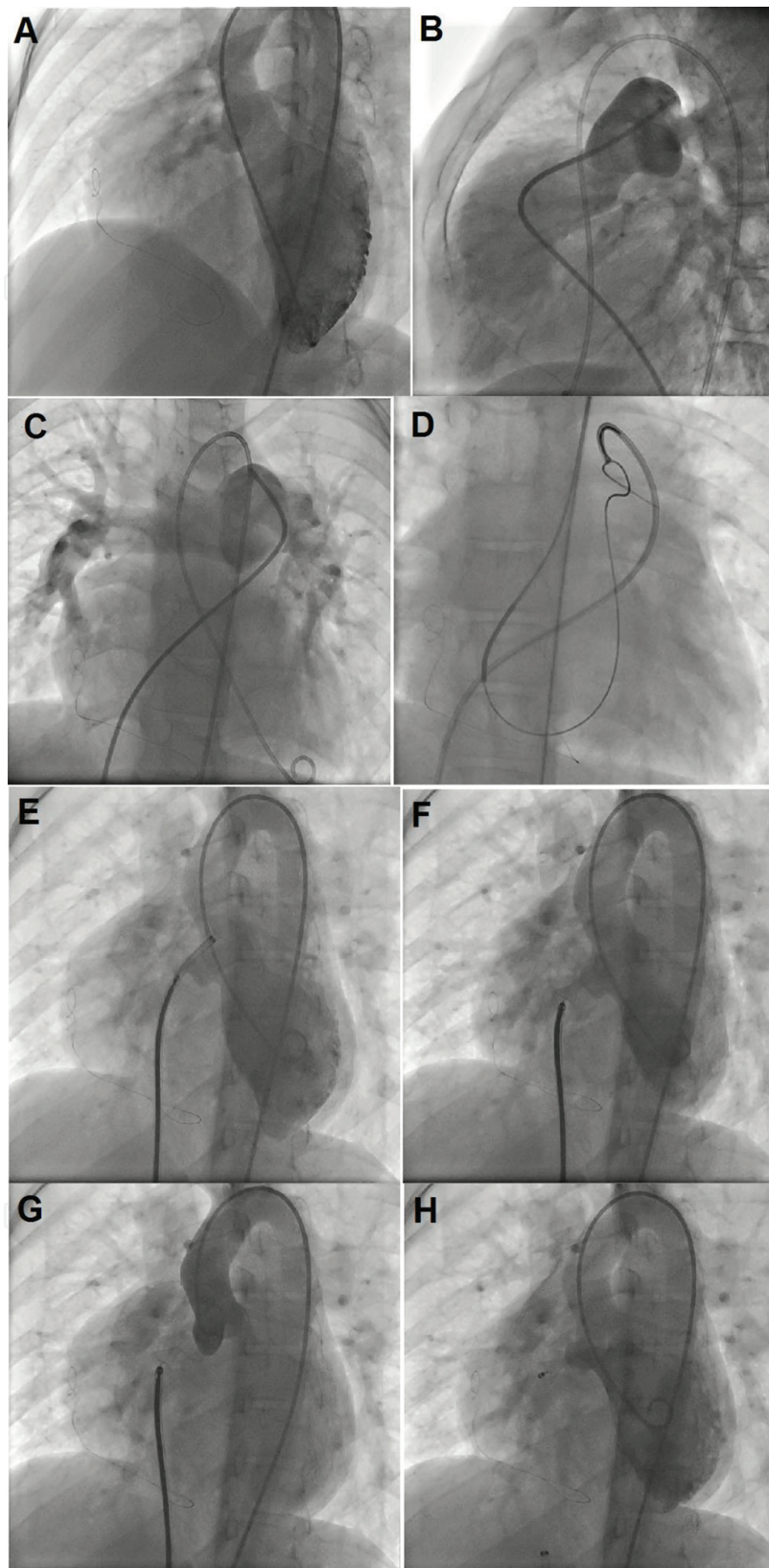


Figure 5. Post operation residual VSD closure technique in a patient with TOF using Occutech Duct Occluder. (A) Small residual VSD around the patch. (B) Main pulmonary injection showing free PI (lateral view). (C) Main pulmonary injection showing free PI (AP view). (D) Snaring the pulmonary wire via venous access making the AV loop. (E) Crossing the VSD into the ascending aorta and releasing the first disc of the device. (F) Pulling back the device and fixing the device to the septum. (G) Aortic root injection, checking the device position and probable AI. (H) Releasing the device.

- Early postprocedure care: three doses of an antibiotic should be given to the patient within the 24 hours of observation with ECG monitoring. Endocarditis prophylaxis and a low dose of an anti-platelet agent like aspirin are recommended for 6 months [4].

5.2. Complications

The complications may occur immediately after the procedure or late during the follow up.

Device embolization: both systemic and right heart embolization of the device may occur and have been reported in up to 2% of cases. Continuous TEE or intermittent TTE during the procedure and especially before the releasing of the device is crucial and very helpful. Operators should be familiar with retrieval techniques, and all necessary equipment for retrieval should be available. Surgical backup is also considered essential [5].

Dysrhythmias: during catheter and device manipulation, temporary dysrhythmias, usually ventricular, are common. Right bundle branch block occurred in only 6% of patients (5) compared to up to 64% of reported surgical series. Complete atrioventricular block (cAVB) is also a potential complication of transcatheter or surgical VSD closure. The reported cases of cAVB are mostly attributed to direct compression trauma, the pressure of radial forces (shape memory of the device), clamping forces, inflammatory processes, and/or the use of oversized devices [6].

Valvular malfunction: the device may influence function of any of the adjacent valves, but especially with the membranous device, aortic and tricuspid regurgitation should actively be looked for with TTE or TEE prior to release of the device.

Hemolysis: blood escape through a small residual shunt after VSD device closure may lead to hemolysis. To be aware of this potential complication especially when there is a residual shunt is very important. Checking the patient's urine color change in early hours after the procedure is a simple way to detect the hemolysis. Some form of little hemolysis may be self-limited and could be managed conservatively but in the form of massive hemolysis blood transfusion and surgical removal of the device should be considered.

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