

Treating Coronary Artery Fistulas and Pulmonary AVMs

Transcatheter coil embolization for coronary and pulmonary arteriovenous malformations.

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Coronary artery fistulas (CAFs) and pulmonary arteriovenous malformations (PAVMs) are relatively rare but clinically important cardiovascular abnormalities that may require percutaneous treatment. In most patients, the current preferred treatment for both lesions is transcatheter embolization using techniques that are well established in these two conditions. We present a review of CAF and PAVM treatment with emphasis on the technical aspects of transcatheter embolization.

TRANSCATHETER TREATMENT OF CAF AND PAVM

Coronary Artery Fistulas

CAFs are rare congenital malformations in which there is a direct vascular connection from a coronary artery to a cardiac chamber, great vein, or pulmonary artery without an intervening capillary bed. CAF is the most com-

mon hemodynamically significant congenital abnormality of the coronary system and is seen in up to 0.22% of adults undergoing coronary angiography. Most fistulas found incidentally are small, single, and result in a left-to-right shunt. In adults, CAF may present as angina, myocardial infarction, or heart failure as a result of a steal phenomenon, with blood bypassing the distal myocardial capillaries (Table 1).

Indications for Treatment and Guideline

Recommendations. Although spontaneous regression has been documented, there are few long-term studies from which to draw conclusions.^{1,2} CAFs have a class II indication for device occlusion therapy. American College of Cardiology/American Heart Association (ACC/AHA) consensus guidelines (based on level of evidence C) indicate that a large CAF, regardless of symptomatology, should be closed via either a transcatheter or surgical

TABLE 1. OVERVIEW OF THE CLINICAL FEATURES OF CAFs AND PAVMs

Clinical Features	CAF	PAVM
Prevalence	1:500 coronary angiograms	15:100,000 general population
Syndrome association	Rare	HHT in 90%
Shunt type	Left to right	Right to left
Common locations	LAD, circumflex, to right heart	Lower lobes, peripheral third of lung
Presenting features	Children: murmur, heart failure Adults: angina, myocardial infarction, heart failure hemopericardium, arrhythmias	Paradoxical embolus (TIA, stroke, cerebral abscess), right heart failure,
Diagnosis	Cardiac angiography	CT/MR/pulmonary angiography
Preferred treatment in appropriate patients.	Transcatheter embolization	Transcatheter embolization

(Courtesy of Cook Medical)

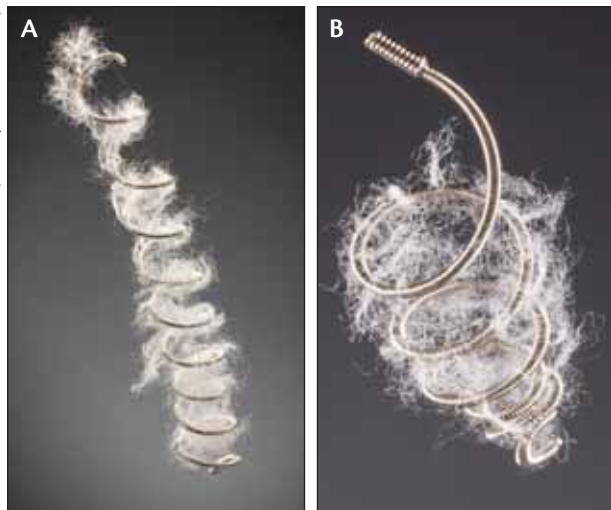


Figure 1. The Nester coil (A). The Tornado coil (B) (Cook Medical, Bloomington, IN).

route after delineation of its course and its potential to fully obliterate the fistula. Furthermore, a small-to-moderate CAF in the presence of documented myocardial ischemia, arrhythmia, otherwise unexplained ventricular systolic or diastolic dysfunction or enlargement, or endarteritis should be closed via either a transcatheter or surgical approach after delineation of its course and its potential to fully obliterate the fistula. Patients with small, asymptomatic CAF should not undergo closure of CAF.³

Small asymptomatic fistulas do not require a specific procedure for closure simply because they are detected incidentally (see *Relative Contraindications to Transcatheter Treatment of CAF* sidebar). However, closure of even small fistulas may be recommended (1) if there is a risk of endocarditis, or (2) if longitudinal follow-up is not feasi-

ble, or (3) if the patient is undergoing an invasive procedure for some other cardiac problem. In children, elective closure of any CAF that remains clinically apparent past 3 to 5 years of age, even if the patient is asymptomatic, is recommended.⁴

Of note in older patients, if the proximal coronary artery has become grossly enlarged, closure of distal fistulas has been associated with a high incidence of late thrombosis of the permanently dilated and tortuous proximal coronary artery.⁵ Closure of the fistula results in relative stasis in the giant coronary artery, and this is likely the etiology of late thrombosis. If the older patient is asymptomatic, the best course of action in this scenario is unclear because late thrombosis may be no better than late development of coronary steal syndrome.

Transcatheter occlusion of a CAF with detachable balloons was first reported by Reidy et al in 1983,⁶ who also reported occlusion of a CAF with coils in 1991.⁷ Because of its minimally invasive nature and high success rate, transcatheter closure with either pushable coils^{4,8-11} or an Amplatzer device (AGA Medical Corporation, Plymouth, MN) is considered the procedure of choice in most patients requiring treatment. In experienced hands, residual leaks are rare (< 5%) and are usually amenable to retreatment. In our experience, the majority of patients are satisfactorily and safely treated with transcatheter coil embolization.

Technique for Coil Placement. Systemic heparin administration (100 units/kg) is recommended after the femoral venous and arterial accesses are established. The choice of catheter system is as important as that of coil type and size in terms of facilitating precise coil placement in fistulas that are often dilated and tortuous. We have used a

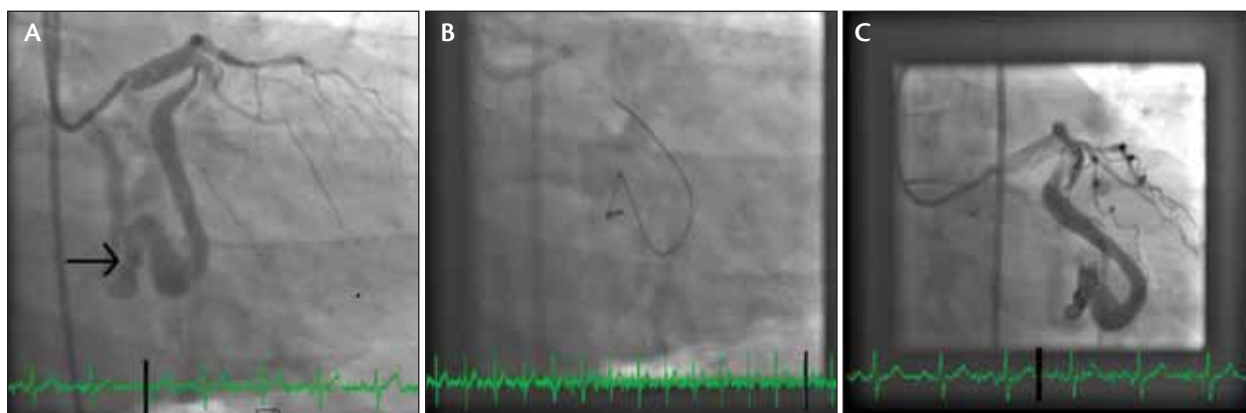


Figure 2. Coronary angiography using an 8-F guide catheter shows a fistula between the circumflex and the right atrium. The proximal coronary dissection is stable (A). Partial deployment of an 0.018-inch Tornado coil via a 3-F catheter advanced through the guide sheath to the narrowest part of the fistula (B). Deployment of more coils results in a reduction of flow in the fistula. Further angiography after 3 minutes showed complete occlusion (C).

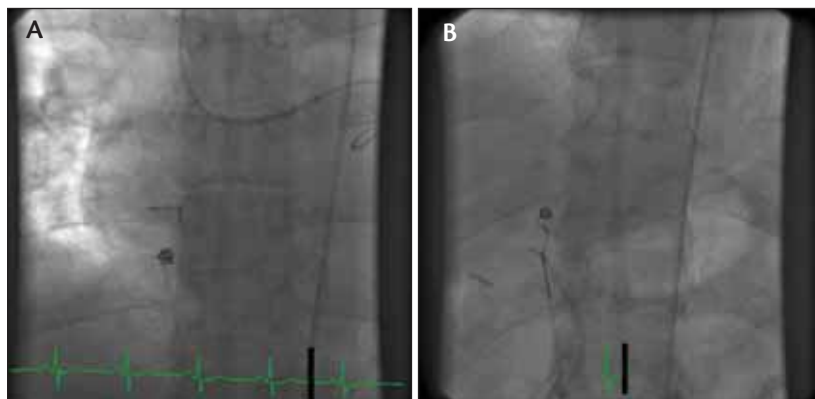


Figure 3. A guidewire has been advanced from the right common femoral vein through the coil, which had migrated through the large fistula. A loop snare has been advanced over the guidewire (A). The loop snare with the coil is withdrawn into its guide catheter and removed (B).

triaxial (3-F/5-F catheters via an 8-F guide catheter) system for more stability and precise placement in tortuous vessels. A coaxial (5-F catheter/8-F guide) system should suffice in less tortuous anatomy. A useful test of catheter tip stability is to advance the coil pusher wire (0.016 inches in the case of microcoils, LLLT [Newton] guidewire for 0.035/0.038-inch coils) to the tip of the catheter first as a trial run to gauge the likelihood of catheter displacement or backing out. We typically use pushable, as opposed to detachable, coils because they are less expensive and easier to stock in a shared interdepartmental inventory. Collaboration with interventional radiologists for choice and deployment of coils is recommended for cardiologists who do not implant coils regularly.

The choice of 0.018-inch versus 0.035-inch coils is dependent on the delivery catheter size, which in turn is dependent on the tortuosity of the fistula. We oversize coil diameter by 20% relative to the CAF diameter. Nester coils are composed of a platinum wire base and allow formation of a more compact coil ball than comparable stainless steel coils (Figure 1A). Tornado coils are made with either the smaller end or larger end (LEF) deployed first (Figure 1B). With the delivery catheter in place, the coil is deployed by using a gentle tapping motion on the coil pusher, which permits visualization and control of the proximal coil end until it exits the catheter. We aim to create a compact coil ball or plug in the fistula because we believe that this promotes more rapid and complete occlusion (Figure 2).

Patience is essential while waiting for cessation of flow after embolization. Angiography performed after embolization may reveal additional smaller fistulas due to change in flow dynamics caused by embolization—which may not require immediate treatment. Placement

of a covered stent across the origin a coronary fistula may be the appropriate choice in adult patients with coexisting coronary artery disease.^{9,10}

Amplatzer Occluders. The main advantage of using an Amplatzer device is speed in deployment. However, its relative bulk and that of its delivery system may prevent its use in smaller and more tortuous fistulas. This challenge is greatly facilitated by initially advancing a guidewire through the fistula, where it is snared on the venous side. This through-and-through access can then be upsized, allowing the use of a larger and more stable platform for precise deployment of

the plug device.^{11,12} Oversizing of the vessel by 30% to 50% is recommended for the current Amplatzer Vascular Plug II (AVP II). Deployment is a combination of unsheathing the device and then unscrewing the delivery wire. Advancement of the unsheathed device is not recommended.

Complications of Embolization. Complications associated with embolization include migration of coils, transient ST-T wave changes,¹³ transient arrhythmias,¹⁴ distal coronary artery spasm, and fistula dissection.⁸ Air embolization may occur, and careful sheath and delivery catheter management is essential. Interventionists should be familiar with the use of loop snares to retrieve coils that have migrated through a fistula (Figure 3).

Surgical Treatment. Up to 30% of patients with CAF have an additional congenital anomaly, most commonly tetralogy of Fallot, patent ductus arteriosus, and atrial septal defect.¹⁵ Surgical closure is the preferred approach in patients who are undergoing operative repair of other cardiovascular problems. The surgical approach can also allow for reduction in the size of very large aneurysmal dilations of either the fistula or the proximal coronary artery.^{16,17}

Follow-Up. Most of the data on long-term coronary patency are based on patients who have had surgical closure of CAF. The long-term patient outcomes after fistula embolization remain unknown, but intermediate-term results reveal persistent coronary artery dilatation in many of these patients.⁵ Because there is little information on the natural history of the patency of aneurysmal coronary arteries after transcatheter embolization, it may be wise to evaluate myocardial perfusion on follow-up.

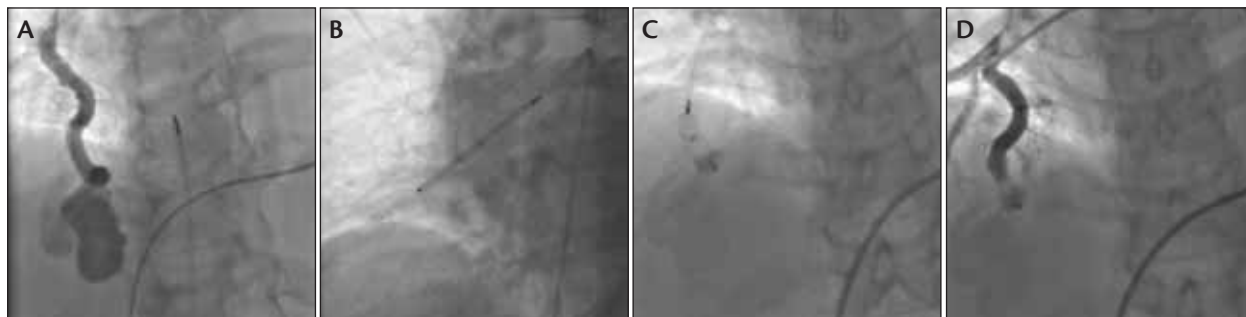


Figure 4. Selective right lower lobe pulmonary angiography shows an AVM with aneurysmal dilatation (A). Deployment of an Amplatzer Plug via the delivery sheath (B). The device has expanded after withdrawal of the sheath (C). Repeat pulmonary angiography after deployment shows complete occlusion of the PAVM by the Amplatzer Plug (D).

Based on limited experience, long-term anticoagulation with at least one antiplatelet agent is suggested in patients with aneurysmal dilatation, and some cardiologists suggest warfarin.^{4,5,18}

Pulmonary AVM

PAVMs are defined as direct, low-pressure, artery-to-vein connections that result in a right-to-left shunt. They are rare, usually congenital abnormalities, with a strong association with hereditary hemorrhagic telangiectasia (HHT), an autosomal dominant disorder in which approximately one-third of patients develop PAVM (Table 1). If untreated, complications of PAVM may occur in up to 50% of patients; these include transient ischemic attacks, paradoxical embolization, massive hemoptysis, or hemothorax. The goals of treatment are threefold: (1) improvement of dyspnea/hypoxemia, (2) prevention of lung hemorrhage, and (3) prevention of neurologic sequelae.

Diagnosis. Initial screening with contrast echocardiography, followed by measurement of PaO₂ while breathing 100% oxygen, is the optimal screening procedure for identifying patients with PAVM, whereas screening with chest radiography and pulse oximetry alone has been shown to be insufficient.^{19,20}

Computed tomography, magnetic resonance imaging, and pulmonary angiography are equivalent in accuracy for the detection and confirmation of PAVM.²¹⁻²³ In patients with HHT, PAVMs were diagnosed as a result of systematic screening procedures (29%), incidental imaging findings (15%), dyspnea (22%), or central nervous system symptoms (13%).

We have previously described a method to diagnose and localize PAVM during a catheterization procedure using intracardiac echo in conjunction with selective injection of bubble contrast into the pulmonary arteries. This allows for the rapid and accurate diagnosis of PAVF

when a patent foramen ovale is sought but not found and for the exclusion of associated PAVF when a patent foramen ovale exists.²⁴

Coils. Pulmonary angiography with measurement of pulmonary arterial pressures is typically done first. The catheter is then exchanged for a coaxial 80-cm, 7-F/100-cm, 5-F catheter system. The use of a guiding catheter avoids elongation of the coil during deployment, resulting in a more tightly packed mass of coils and better cross-sectional occlusion of the vessel. More importantly, guiding catheters provide enhanced stability and control during coil deployment.

Although detachable coils offer a more controlled delivery with a lower risk of coil migration, they are more expensive. White developed a technique of using coaxial catheters to deliver less-expensive pushable coils, which provides safe and controlled deployment. These operators use a 5-F coil delivery catheter within a 7-F guiding catheter. White states, "It is not the coil that is so important, but the use of coaxial or triaxial catheters that allow for precise placement of the coil."²⁵ Cross-sectional occlusion is essential for embolization of PAVM and this is achieved using the anchor or scaffold coil technique. The anchor technique entails deploying the initial segment of coil in an adjacent vessel or side branch as an anchor before deploying the bulk of the coil in or adjacent to the PAVM itself.²⁵

White also recommends that guidewires always be withdrawn from catheters under saline flush to prevent air embolism via the catheter. In addition, regular flushing of catheters and systemic periprocedural heparinization to reduce the potential for embolization of particulate matter or clot is advocated.²⁵

Amplatzer Device. The AVP II is a redesigned version of the original AVP, with the important modifications of having a finer, more densely woven nitinol frame and also a multisegmented design. The device is available in sizes

RELATIVE CONTRAINDICATIONS TO TRANSCATHETER TREATMENT OF CAF

- Size is too small
- Extreme tortuosity preventing delivery of a catheter far enough distally for embolization
- Presence of multiple fistula
- Proximity of normal coronary branches to CAF
- Grossly enlarged proximal coronary associated with a distal fistula

ranging from 4 to 22 mm in 2-mm increments, and can be delivered through guide sheaths between 4 and 7 F, depending on the device size. The Amplatzer device produces almost immediate complete closure, and recanalization has not been reported. The device is available in a range of five sizes, from the 4-6-mm device for small vessels to the 12-14-mm device for large vessels, and requires delivery through 5- to 7-F sheaths, allowing its use in small infants and neonates.²⁶ Multiple devices can be delivered through a single sheath, and the device can be repositioned or retrieved until the operator is sure of proper positioning. A disadvantage of the device may be its relatively long length, which may preclude its use if the target vessel is short (Figure 4).

Hart reported successful embolization in 75% of 161 PAVMs using AVP alone. Complete and rapid occlusion of feeding vessels was easily achieved at the site of arteriovenous communication without complication. Particularly small or tortuous feeding arteries supplying 27 complex and 14 simple PAVMs were occluded with coils. There have been no documented instances of recanalization on follow-up.²⁷

Detachable Occlusion Balloons. Before their removal from the United States market in 2002, detachable latex and silicone occlusion balloons were a useful and effective method of treating PAVMs. Follow-up studies showed a low recanalization rate despite early balloon deflation.

Complications. Complications of transcatheter PAVM embolization include infections, lung infarction, pleural pain and effusion, air embolism, cardiac arrhythmia, transient angina, and paradoxical embolization with cerebral infarction.²⁸⁻³¹ Felix et al describe a patient who had a stroke immediately after embolization treatment of a PAVM and recommend antiplatelet drug use, such as low-dose aspirin, as the short-term primary stroke prevention after endovascular PAVM embolization.³²

Outcomes. Trerotola et al report that outpatient single-session embolization was achieved in 44 of 51 patients

(86%), with most patients discharged after a 2-hour period.³³ Recurrence is rare (< 2%), with recanalization being the most common mechanism of PAVM reperfusion accounting for 88% of cases.³⁴ Increased feeding artery diameter, low number of coils, use of oversized coils, and proximal coil placement within the feeding artery are associated with reperfusion. Distal coil placement facilitates repeat embolization if reperfusion occurs. Repeat embolotherapy for reperfused PAVMs was technically successful in 94% of cases. In the remaining 6% of cases, insufficient feeding artery length prevented safe repeat treatment.³⁵

Follow-Up. Clinical and anatomic evaluation after PAVM embolization is important to detect persistent or reperfused lesions and enlarging lesions, with the latter more common. Patients with persistent, reperfused, or enlarging lesions often have symptoms, but many patients may be asymptomatic. More frequent assessment, such as annual pulse oximetry, contrast echocardiography, and chest radiography have been suggested to improve detection before the onset of symptoms.^{26,34}

All patients should be assessed for other manifestations of HHT before treatment, and they are best followed in one of the 20 HHT centers worldwide (www.hht.org).

SUMMARY

Transcatheter embolization of CAF and PAVM is effective in reducing both anatomic shunt fractions and in reducing complications. The technique of transcatheter embolization is well tolerated and has a low complication rate. Collaboration with interventional radiology/neuroradiology for choice and deployment of devices is recommended. ■

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