

Pulmonary Arteriovenous Malformations and Coronary Artery Fistulas

Diagnosis, evaluation, and indications for closure.

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Pulmonary arteriovenous malformations (PAVMs) and coronary artery fistulas (CAFs) are infrequent clinical problems but can potentially cause significant morbidity and mortality. They are usually congenital, but acquired forms do occur. Treatment of these vascular abnormalities has evolved considerably during the last 2 decades. Percutaneous transcatheter closure of PAVMs and CAFs is now considered to be an effective and safe alternative to surgery. This article summarizes the evaluation and indications for closure in patients diagnosed with these vascular abnormalities.

PAVMs

Diagnosis

PAVMs consist of abnormal direct, low-pressure communication between the pulmonary arteries and veins. Since the first reported case in 1897, more than 500 cases have been reported.¹ Most PAVMs are congenital, of which, 60% to 90% are associated with hereditary hemorrhagic telangiectasia (HHT).² Acquired causes of PAVMs include mitral stenosis, thoracic surgery, hepatic cirrhosis, trauma, tuberculosis, schistosomiasis, and actinomycosis.³ They can also be seen in patients with palliated complex congenital heart disease (ie, post-classic Glenn shunt). PAVMs result in a right-to-left shunt, which may cause symptoms. Because the normal filtering function of the lung is impaired in patients with PAVMs, they can also be a source of paradoxical embolism and infection, resulting in strokes, transient ischemic attacks, and cerebral abscesses. Table 1 lists the complications.

PAVMs occur twice as often in women as in men. If symptoms develop, they often occur between the ages of 40 and 60 years. Clinical features may include epistaxis (in patients with HHT), dyspnea, hemoptysis, cyanosis, and

clubbing (in the presence of a large right-to-left shunt). In the majority of cases, patients with PAVMs have an underlying genetic disorder of the blood vessels, HHT. The most commonly reported complications relate to the central nervous system, resulting from right-to-left shunts or coexisting cerebral AVM, with the incidence being 19% to 59%.^{4,5} Such patients may experience brain abscess or stroke.

Evaluation

Approximately 53% to 70% of PAVMs are seen in the lower lobes, with the left lower lobe being the most common location.^{6,7} They are found in close proximity to the visceral pleura or embedded in the outer third of lung parenchyma. Because most PAVMs are located at the bases of the lung, platypnea (improvement in dyspnea when lying down) orthodeoxia (desaturation while upright) may be a presenting symptom.

TABLE 1. COMPLICATIONS OF PAVMs

Neurological

- Cerebral abscess
- Cerebrovascular accident
- Transient ischemic attack
- Migraine
- Seizure

Cardiovascular

- Pulmonary hypertension
- High-output cardiac failure
- Paradoxical embolism

Pulmonary

- Hemoptysis
- Hypoxemia/orthodeoxia
- Hemothorax

Hematological

- Polycythemia

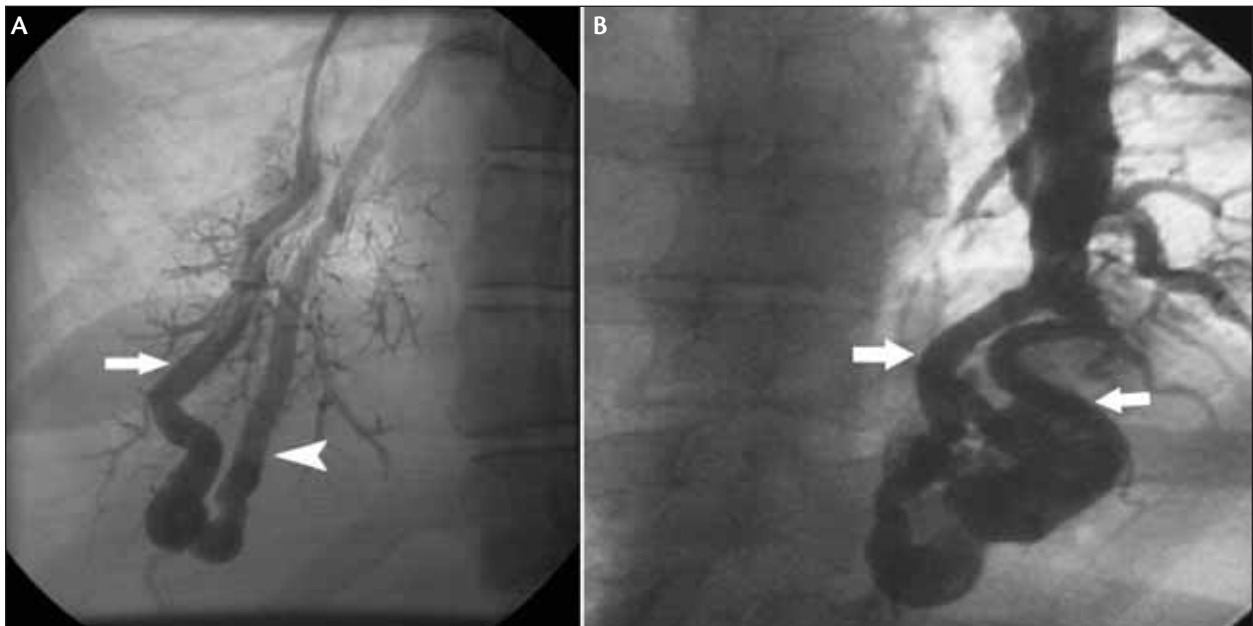


Figure 1. Pulmonary angiography demonstrating a simple PAVM with one feeding artery (white arrow) and one draining vein (white arrowhead) (A). Pulmonary angiography of a complex PAVM demonstrating two feeding arteries (white arrows) (B).

PAVMs have been classified as simple, complex, or diffuse depending on the number of feeding arteries and veins. Most PAVMs (80%–90%) are the simple type, which is defined as those with a single segmental feeding artery and draining vein. The complex type is defined as having two or more arteries supplying the PAVM and one or two draining veins (Figure 1A, B). There is often an aneurysmal sac connecting the artery and vein. Diffuse PAVMs are rare and consist of multiple small PAVMs throughout the lung.

A variety of diagnostic modalities can be utilized to evaluate PAVMs. Chest radiography is an important and readily available diagnostic tool because 90% of PAVMs are seen on a chest radiograph.⁸ Their classic radiographic features are a poorly defined round or oval mass of uniform density that is frequently lobulated (ranging in size from 1 to 5 cm in diameter) and found more commonly in lower lobes.

Contrast-enhanced computed tomography (CT) scanning is a valuable tool in diagnosis and in defining the vascular anatomy of PAVM. CT scanning is more sensitive than conventional pulmonary angiography in detecting PAVMs (98% vs 60%).⁹ The superiority of CT scanning in detecting PAVMs is attributed to the absence of superimposition of the lesion in transaxial views (Figure 2). However, CT scanning may underestimate the size and importance of the lesions. Pulmonary angiography is better in determining the angioarchitecture when supplemented with subselective angiograms. Pulmonary angio-



Figure 2. Axial view of a contrast-enhanced CT scan demonstrating a PAVM (white arrow).

graphy remains the gold standard in the diagnosis of PAVM and is necessary if resection or embolization is being considered.

Contrast echocardiography is a sensitive tool in the detection of small right-to-left shunts. This technique involves injecting echocardiographic contrast, such as agitated saline, into a peripheral vein while simultaneously imaging the right and left atria with 2D echocardiography. In the case of PAVM and other anatomic intrapulmonary shunts, there is nearly always a delay of greater than four cardiac cycles before contrast is visualized in the left atrium, whereas with intracardiac shunts,

the contrast is visualized in the left heart chambers within three to four cardiac cycles of its appearance in the right atrium. This technique can sometimes be too sensitive by detecting clinically insignificant microvascular shunts.

Other adjunctive, highly sensitive diagnostic techniques include radionuclide perfusion scanning and shunt fraction measurement.^{4,10} However, we do not routinely use them in clinical practice because they are unable to differentiate between an intracardiac versus a pulmonary source of shunt. Magnetic resonance imaging (MRI) is not commonly used because of the expense and availability.

Indications for Closure

Although the natural history of untreated PAVM has not been optimally delineated, it is clear there is considerable morbidity and mortality in some patients. In general, indications for treatment include three broad categories: exercise intolerance, prevention of neurologic complications, and prevention of lung hemorrhage. The most common indication for occlusion of PAVMs is to prevent neurologic complications. Serious neurologic events, including transient ischemic attack, stroke, and brain abscess, occur in up to 30% to 40% of patients with PAVMs that have feeding arteries 3 mm or greater in diameter.^{8,11} In addition, these larger PAVMs are more prone to rupture. In 80% of cases, patients with large PAVMs have smaller arteries present as well. Although not candidates for embolization, these smaller malformations still pose a risk for infection, air emboli, and brain abscess. Therefore, it is important that patients return for clinical follow-up as well as CT imaging 1 year after occlusion of the larger PAVMs. This allows for confirmation that the treated malformations remain occluded and also enables monitoring of untreated, smaller malformations.¹¹ Any evidence of persistence suggests recanalization and may be an indication for re-embolization.

Since the late 1990s, transcatheter embolization has replaced surgery as the treatment of choice. A number of different materials have been used to embolize PAVMs, including various types of fibered and unfibered, detachable and pushable coils, and most recently, the Amplatzer vascular plug (AGA Medical Corporation, Plymouth, MN). The techniques, advantages, and disadvantages of these embolotherapies are beyond the scope of this article.

Due to the considerable morbidity and mortality of PAVMs without treatment, all patients who are symptomatic due to PAVMs and patients with feeding arteries that exceed 3 mm in diameter, should be treated with embolotherapy to potentially limit serious complications.

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Also, surveillance of occluded PAVMs is mandatory to detect recanalization and growth of small, unoccluded PAVMs that can frequently enlarge with time.

CAFs

Diagnosis

CAFs are congenital or acquired coronary artery abnormalities in which blood is shunted into a cardiac chamber, great vessel, or other vascular structure, bypassing the myocardial capillary network. CAFs are usually congenital in origin and appear to represent the persistence of embryonic intertrabecular spaces and sinusoids.^{12,13} They may be present as solitary fistulas or coronary artery–left ventricular multiple microfistulas (CA-LVMMFs). The incidence of CAFs is estimated to be 0.02% to 2.1% (2.1% represents CA-LVMMFs) in patients undergoing coronary arteriography.^{14,15} They can occur from any of the three major coronary arteries, including the left main trunk. CAFs arise from the right coronary artery (RCA) in approximately 55% of patients, from the left coronary artery (LCA) in 45% of patients, and from both the RCA and LCA in approximately 5% of patients.¹⁶ The majority of symptomatic CAFs originate from the RCA.¹⁷ More than 90% of these fistulas drain into a low-pressure, right heart structure.¹⁸ Fistulous drainage in adults most commonly involves the pulmonary artery (57%), followed by the right atrium (12%), coronary sinus (12%), and right ventricle (10%). However, in children, the drainage most often is seen into the right ventricle (50%), followed by the pulmonary artery (32%) and the right atrium (31%). Fistulous communication to the left-sided chambers is less frequent (6%–10%).¹⁹

The diagnosis of a CAF is often suspected on auscultation when a continuous murmur is noted; although, the interventionist must consider alternative diagnoses such as ruptured sinus of valsalva aneurysm, patent ductus arteriosus, internal mammary artery to pulmonary artery fistula, and systemic arteriovenous fistulas. The clinical presentation is variable in both the adult and pediatric populations. Newborns can present with signs and symptoms of congestive heart failure immediately after birth due to the increased volume load on the left heart. Most pediatric patients, however, are asymptomatic at the time of diagnosis. Although only 10% of pediatric patients will

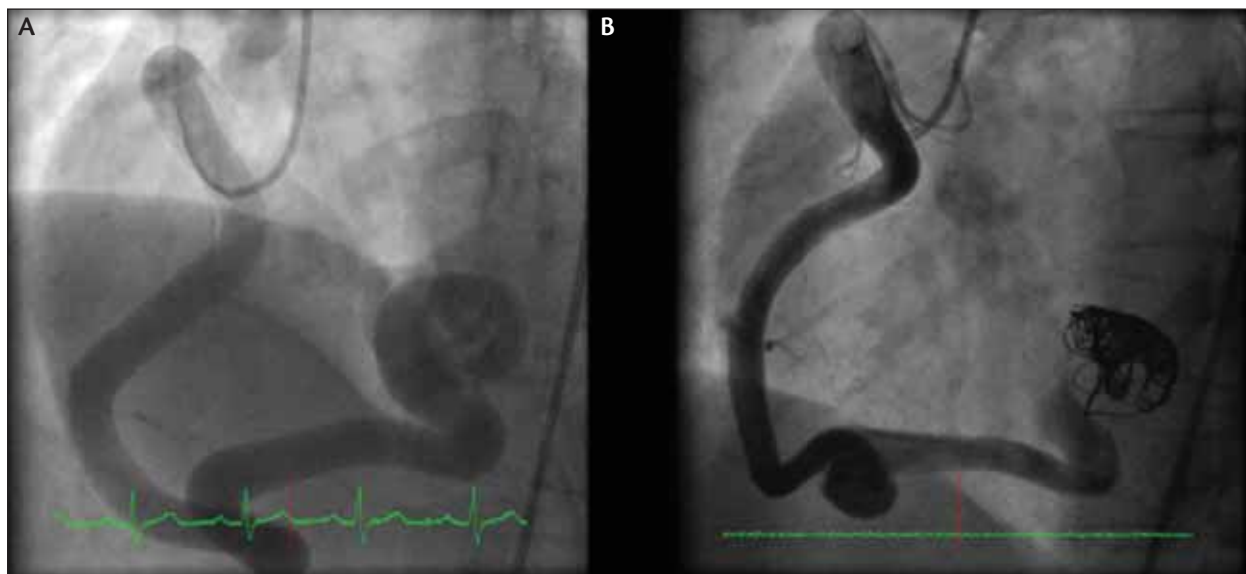


Figure 3. Coronary angiography demonstrating an RCA to coronary sinus fistula (A). Coronary angiography 2 months after successful coil embolization demonstrating minimal residual flow across the RCA to coronary sinus fistula (B).

be symptomatic, adults are much more likely to report symptoms. The incidence of symptoms in adults has been reported to be up to 79%.¹⁴ In adults, dyspnea and chest pain are the most frequently encountered symptoms. Symptoms of angina may, in part, be related to coronary steal, with blood preferentially flowing to the lower pressure chamber. Other potential complications include endocarditis, arrhythmias, spontaneous rupture, and thrombosis resulting in myocardial infarction. Rarely, the presenting feature can be pericardial effusion or sudden death.

Evaluation

Many fistulas are small and are found incidentally during coronary arteriography, which is the gold standard for evaluation of CAFs. Coronary arteriography reliably demonstrates the proximal part of the CAF and allows the evaluation of the size, its relationship to the coronary arteries, and their derivative branches. The interventionist should also evaluate for the presence of concomitant atherosclerosis and other structural abnormalities at the time of cardiac catheterization. A complete hemodynamic evaluation should be performed to document the magnitude of the shunt and to evaluate for any potential complications, such as pulmonary hypertension. A net left-to-right shunt usually exists, however, the shunt ratio is generally small, regardless of patient age.

A markedly enlarged coronary artery can usually be detected by transthoracic, 2D echocardiography. This is more successful in children in whom optimal acoustic

windows are obtained. Continuous turbulent systolic and diastolic flow pattern characterizes the shunt entry site. Use of agitated saline contrast to enhance the color Doppler signals can assist in defining the location and extent of CAFs.

Other imaging modalities have proven useful for characterization of CAFs. Contrast-enhanced CT is a useful, noninvasive, and accurate imaging technique for the detection of CAFs. Multidetector CT has been shown to provide a high-resolution anatomic image by using electrocardiogram-gated reconstruction methods because it allows for evaluation of aneurysmal dilatation or thrombus formation in the vessel. Volume-rendered images acquired from 3D CT data sets provide an excellent overview of the cardiac and vascular anatomy and help the interventionist or surgeon understand the anatomical complexity. Submillimeter reconstruction gives multidetector CT higher temporal and spatial resolution than those of MRI. The major limitation of CT is radiation exposure.²⁰

Before closure, precise anatomic delineation of origin, tortuosity, termination, branch points, caliber, and tapering of the fistula is critical to procedural planning and estimation of procedural success and equipment choice. Atypical symptoms should be scrutinized, and an effort should be made to evaluate for chamber enlargement, inducible ischemia, and wall motion abnormalities by way of noninvasive testing.

Indications for Closure

Spontaneous closure secondary to thrombosis or atherosclerosis, although rare, has been reported. The inci-

dence of spontaneous closure in all age groups is estimated at 1% to 2% and is more common in the pediatric age group.¹⁴ If the fistula is small, the overall prognosis is good for both the adult and asymptomatic pediatric population. These small CAFs do not require intervention and only sporadic follow-up is necessary.

Management options include surgical ligation, percutaneous transcatheter embolization (PTE), watchful waiting, and medical treatment. Although in the past the traditional approach was surgical closure, most CAFs are now being closed in the catheterization laboratory. This can be performed with the use of coils, vascular occlusion devices, and Amplatzer occluders of various morphologies. Features in favor of PTE are proximal location of the fistulous vessel, extra-anatomic termination of the fistulous vessel away from the normal coronary arteries, and absence of concomitant cardiac disorders requiring surgical intervention.

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The decision to close the fistula using PTE should be based on multiple factors. These include symptoms, fistula size, magnitude of shunt, fistula origin and termination, number of feeding vessels, risk to viable myocardium, risk of residual coronary artery thrombosis, and likelihood of success. Moderate or larger fistulas that are symptomatic cause draining chamber enlargement, are larger than the ongoing coronary artery, or are aneurysmal, should most likely be treated. The aim of the catheter procedure should be to achieve complete occlusion at as distal a location as possible in the fistulous vessel (Figure 3A, B).

The American Heart Association guidelines for the management of adults with CAFs have recommended the following: (1) a large CAF, regardless of symptomatology, should be closed via either a transcatheter or surgical route after delineation of its course and its potential to fully obliterate the fistula, and (2) 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.

CONCLUSION

The natural history of patients with CAFs is greatly variable. Many patients remain asymptomatic throughout life. Patients who have had successful closure of CAFs require long-term antiplatelet or anticoagulant therapy because of the potential risk of coronary thrombosis. Long-term follow-up is essential due to the possibility of recanalization, persistent dilatation of the coronary artery, thrombus formation, and myocardial ischemia. ■

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