Endovascular Repair of PAVM by Embolotherapy

In a patient with a transient ischemic attack or stroke, a positive contrast echocardiogram does not always mean a patent foramen ovale.

BY ROBERT I. WHITE, JR, MD

Pulmonary arteriovenous malformations (PAVMs) are a direct communication between the pulmonary artery and vein, via a thin-walled aneurysmal sac and often go undetected. A patient presenting to the emergency room with symptoms of transient ischemic attack (TIA) or stroke undergoes CT to exclude cerebral hemorrhage. Often, the next test(s) will include carotid ultrasound, as well as contrast echocardiography (CE) to exclude an embolic origin for the neurologic symptoms. If the results of the CE are positive, with microcavitations appearing within the left atrium within three to six cardiac cycles, a diagnosis of right-to-left shunting is established. The embolic stroke is then attributed to an embolus crossing a patent foramen ovale (PFO), and the patient may undergo attempted PFO closure by transcatheter techniques. PAVMs should also be included in the differential diagnosis of a positive CE in a patient with neurologic symptoms. In this instance, the embolic stroke is due to passage of an embolus through the PAVM rather than a PFO.

If left misdiagnosed or untreated, 50% of patients with PAVM will develop TIA/stroke, brain abscess, or massive hemoptysis or hemothorax.[1,2] Ninety percent of patients with PAVM have an underlying genetic disorder known as *hereditary hemorrhagic telangiectasia* (HHT).[3,4] During the evaluation of a neurologic event, questioning of patients or their families about familial nosebleeds is essential and often overlooked. Forty percent of patients with HHT have either a PAVM or cerebral arteriovenous malformation (CAVM) and may be relatively asymptomatic before their first neurologic event.[2,3] Appropriate attention is now being directed by emergency room physicians toward PFO as a cause of neurologic symptoms, but there is still need for physicians to be more aware of PAVM as a cause for these symptoms in the presence of positive CE results.

HHT is a relatively common autosomal dominant disorder occurring in 200 out of 1 million people.[3] If the patient has a history of nosebleeds, and there is also a positive familial history of epistaxis as well as telangiectases of the oropharynx or fingers, then HHT is probable.[4] Consensus guidelines from the nonprofit organization for HHT (www.hht.org) suggest that all patients with HHT should be screened at least once for PAVM and CAVM.

EMBOLOTHERAPY OF PAVM

During the past 30 years, therapeutic embolization of PAVMs has largely replaced surgical resection.[5-10] Short- and long-term results after occlusion of the feeding artery to the PAVM using pushable fibered coils, and more rarely detachable coils, are excellent when performed by experienced endovascular specialists.[5-10] Recurrences due to recanalization or missed branches occur in less than 10% of patients, and paradoxical embolization of devices rarely occurs when the treating physician is experienced.[5-10] Continued surveillance of patients is required minimally at 5-year intervals to detect growth of unoccluded PAVMs, which occurs in up to 20% of patients.[8,10] Because stroke or TIAs are twice as common in patients with multiple as opposed to single PAVMs with 3-mm-diameter feeding arteries, we advocate closure of all PAVMs with feeding arteries 3 mm or greater in diameter.[11]
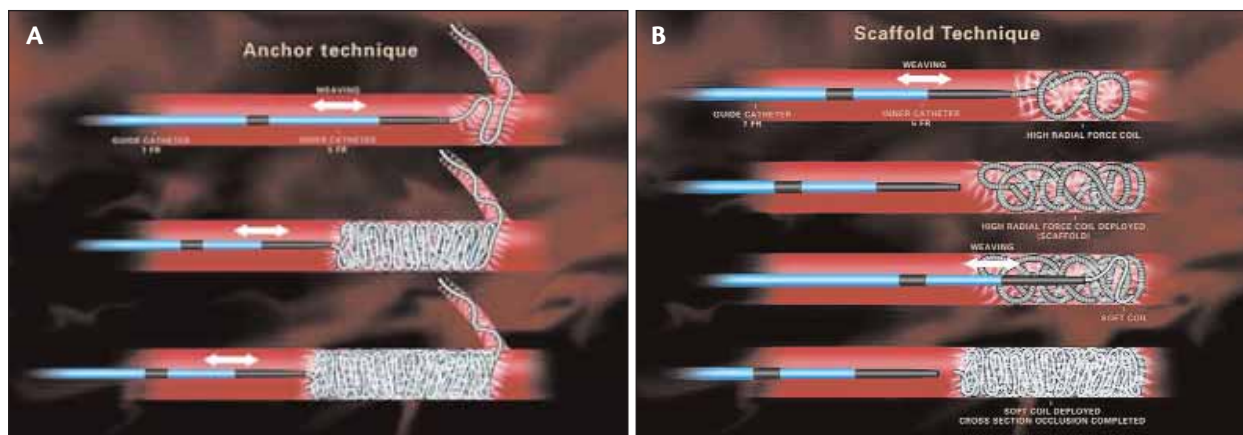
Figure 1. Anchor and Scaffold techniques using coaxial catheters and pushable fibered coils. Anchor technique (A): the 7-F guide catheter is placed in the segmental artery to the PAVM. The inner 5-F catheter is hydrophilic and, with small hand injections of contrast material, the catheter is wedged into a side branch close to the aneurysmal sac. The first 2 cm of the fibered coil are anchored into the side branch, and the remainder of the coil is packed into a tight coil nest. The addition of one or two more tightly packed coils results in cross-sectional occlusion. Scaffold technique (B): in large PAVMs, oversized high-radial-force coils (.035-inch) are deployed proximally to the aneurysmal sac. They may also be anchored if there is a suitable vessel adjacent to the sac. Precision and safety are provided by the guide catheter held constant, proximally in the segmental artery to the PAVM while deploying the coil through the 5-F inner catheter. After placement of one or two MReye coils, softer fibered platinum coils are nested within the scaffold to complete the occlusion. (Adapted from White RI, Pollak JS. Controlled delivery of pushable fibered coils for large-vessel embolotherapy. In: Springer, Ed. Vascular embolotherapy: a comprehensive approach. Berlin, Heidelberg, New York: Jafar Golzarian. 2006;35-42.)

PAVMs are classified as simple or complex, depending upon whether there is a single segmental branch (simple) or multiple segmental branches (complex) connecting to the aneurysmal sac and draining vein. While standard CT angiography or MR angiography may be all that is required for classifying the type of PAVM, they are not a substitute for pulmonary angiography, including selective injections with the guiding catheter in the segmental artery to the PAVM. A segmental injection in the feeding artery in multiple projections will be the most important part of any diagnostic angiogram. By determining the anatomy of the artery as it enters the sac, the method of closure is decided upon.

Pulmonary angiography should be performed with "intent to treat." This includes selective injections in the right and left pulmonary artery in multiple projections. It is our preference to treat one lung at a time because 15% of patients develop delayed self-limited pleurisy occurring 2 to 7 days after treatment and due, in most instances, to thrombosis of a pleural-based sac. Our standard approach is percutaneous from either femoral vein, leaving a 7-F sheath in the femoral vein attached to a heparinized flush. Several precautions are followed during therapy, which has led to fewer complications. Patients are heparinized with a single dose of heparin (100 units/kg). Fastidious measures to avoid any inadvertent air delivered through the catheter are fol-

lowed. Because these patients have right-to-left shunting, small amounts of air passing through the PAVM can lead to air emboli in the coronary or carotid arteries. All guidewires are removed slowly under saline in a wide bowl to avoid a vacuum effect, and no catheters are flushed without good blood withdrawal.

TECHNICAL ASPECTS OF EMBOLOTHERAPY

Most specialists are familiar with pushable fibered coils, and they have been our standard method for closure during the past 7 years. The Amplatzer Plug (AGA Medical Corporation, Plymouth, MN) has been recently introduced but, as yet there are no long-term data on its use for closing PAVMs, and it is a relatively expensive device.[12] More important than the device for closure is the use of a coaxial guide system with the shorter guide catheter placed deep within the segmental artery. We prefer the 80-cm Lumax soft tip guide catheter (Cook Medical, Bloomington, IN) and the 100-cm, 5-F slip catheter (Cook Medical) to deliver the pushable fibered coils for arteries 6 mm or larger in diameter. With smaller arteries from 3 to 6 mm in diameter, we prefer the 5-F angled J catheter (Terumo Medical Corporation, Somerset, NJ) for placing pushable fibered coils. The coaxial technique prevents coil elongation, which was a common cause of recanalization, when pushable fibered coils were placed with a single catheter (Figure 1).
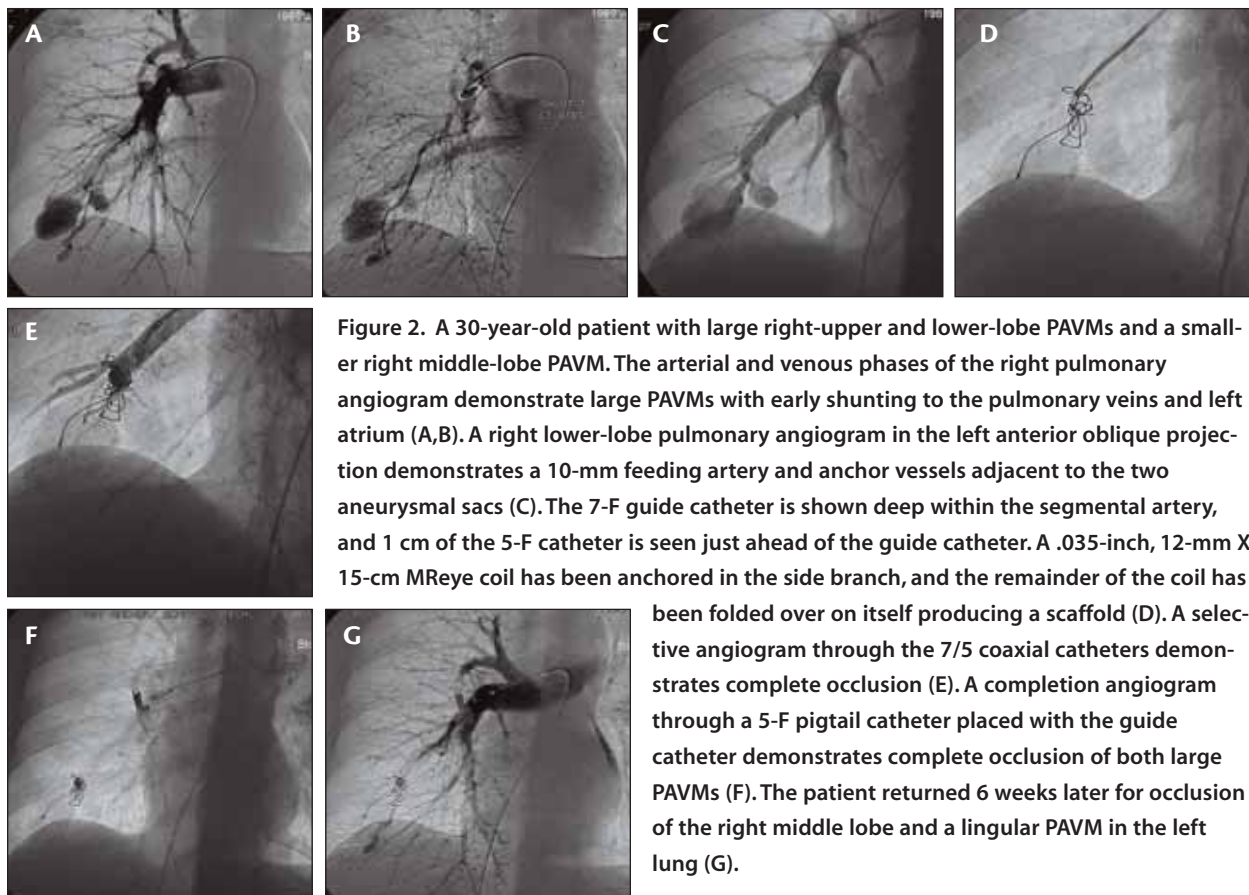
Figure 2. A 30-year-old patient with large right-upper and lower-lobe PAVMs and a smaller right middle-lobe PAVM. The arterial and venous phases of the right pulmonary angiogram demonstrate large PAVMs with early shunting to the pulmonary veins and left atrium (A,B). A right lower-lobe pulmonary angiogram in the left anterior oblique projection demonstrates a 10-mm feeding artery and anchor vessels adjacent to the two aneurysmal sacs (C). The 7-F guide catheter is shown deep within the segmental artery, and 1 cm of the 5-F catheter is seen just ahead of the guide catheter. A .035-inch, 12-mm X 15-cm MReye coil has been anchored in the side branch, and the remainder of the coil has been folded over on itself producing a scaffold (D). A selective angiogram through the 7/5 coaxial catheters demonstrates complete occlusion (E). A completion angiogram through a 5-F pigtail catheter placed with the guide catheter demonstrates complete occlusion of both large PAVMs (F). The patient returned 6 weeks later for occlusion of the right middle lobe and a lingular PAVM in the left lung (G).

Anchor Technique

The Anchor technique (Figures 1A and 2) is utilized to treat most PAVMs, and we have never had paradoxical embolization of pushable fibered coils when this technique is employed.[10] Figure 2 illustrates the Anchor technique in a patient with a right lower-lobe PAVM. The first coil placed always has a diameter 1 to 2 mm larger than the feeding artery, and the first 2 cm of the coil is anchored in a side branch close to the aneurysmal sac. Anchoring the first coil prevents paradoxical embolization. Key to the use of pushable fibered coils is obtaining cross-sectional occlusion of the feeding artery by packing one to two additional coils to form a tightly nested coil mass. Coaxial catheters prevent coil elongation routinely experienced in any circulation when placing coils through a single catheter rather than by the coaxial method. If the feeding artery narrows before entering the aneurysmal sac, the anchor technique is rarely used, and we simply place an oversized pushable fibered coil as the first coil with one or two other coils as needed to provide a safe and cost-effective method.

Scaffold Technique

When the feeding artery to the PAVM exceeds 8 mm in diameter and there is no suitable anchor vessel, we have deployed as the first coil, a high-radial-force coil, which is purposely oversized and is long enough to form three loops in the artery (Figures 1B and 3). Again, it is the "purchase" provided by the 7-F guide catheter that allows for precise and safe deployment. These high-radial-force coils originally were made of stainless steel and have now been replaced by oversized and long MReye inconnel coils (Cook Medical). The .035-inch MReye coils have high radial force, much like a self-expanding stent and are prevented from passing through the fistula by their expansion against the initma of the artery. In large PAVMs, if there is an anchor vessel present, the .035-inch MReye coil is anchored as well (Figure 3).

Once the scaffold is formed with one or two MReye coils, soft long-fibered platinum coils are placed within the scaffold and nested into a tight coil mass, producing cross-sectional occlusion. Usually, large fistulae with arteries 8 mm in diameter or larger are effectively occluded with four to five coils. Before finishing one lung, the guide catheter is withdrawn, and the 100-cm, 5-F pigtail catheter is reinserted to do a completion pulmonary angiogram on the side of the freshly occluded PAVM(s).
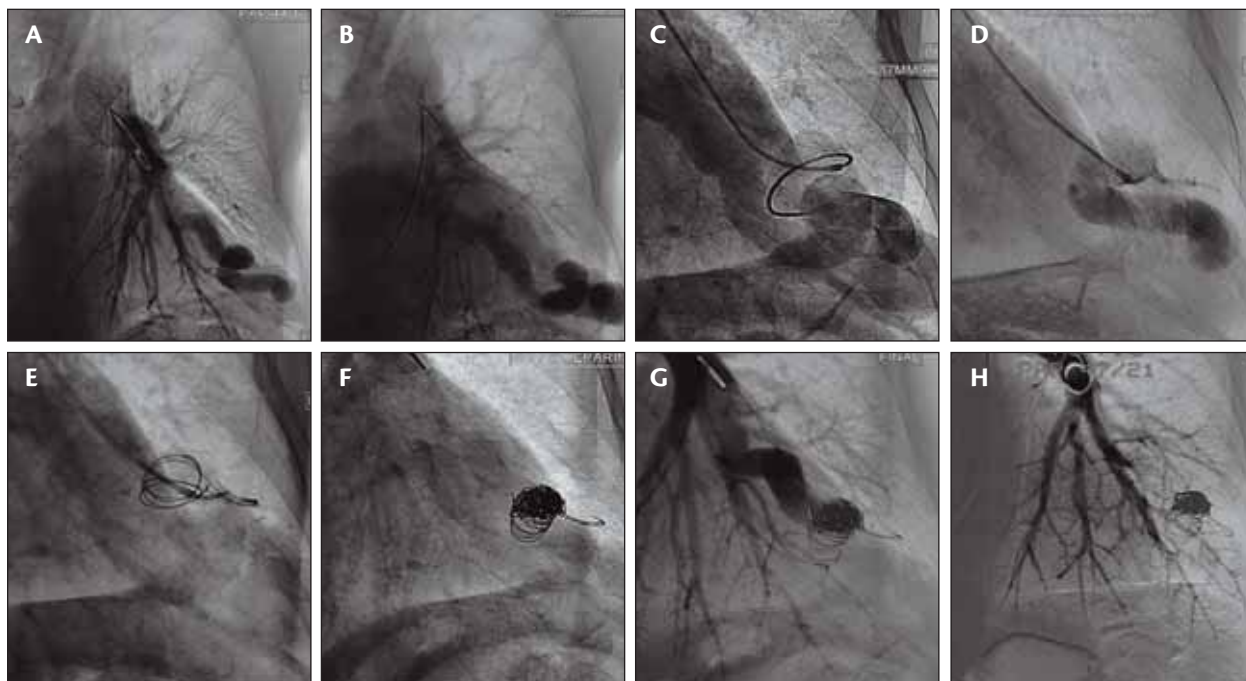
Figure 3. An older patient with mild pulmonary hypertension due to liver AVMs and large left lower-lobe PAVM with a feeding artery measuring 17 mm in diameter. Arterial and venous phases of selective left pulmonary angiogram demonstrate large artery and vein with tortuous aneurysmal sac (A,B). A hand injection of contrast material demonstrates a good-caliber anchor vessel nest to the sac (C). Two 20-mm-diameter X 15-cm-long MReye coils have been anchored in the feeding artery and coiled into a scaffold (D). Fibered platinum coils (12, 10, and 8 mm long) have been packed into the scaffold (E). Left pulmonary angiograms obtained immediately after and 5 months later show no filling of the PAVM (E,F).

Other Technical Considerations

Catheterization of the right middle lobe or lingula may be difficult with the standard 5-F slip catheter or the angled J catheter. As a result, we have utilized the Judkins JL 3.5 100-cm diagnostic catheter (Cordis Corporation, Warren, NJ), placed through the 80-cm, 7-F Lumax guide catheter to access these vessels. By placing the guide catheter in the lower lobe and then placing the JL 3.5 into the guide catheter, it is relatively simple to withdraw the guide and JL 3.5 catheter rotating clockwise and injecting small amounts of contrast material. The 5-F left coronary catheter almost always enters the right middle lobe or lingula and is then advanced into the artery, and the guide catheter is advanced over it. At this point, when dealing with a large fistula, the coronary catheter is removed, and the 5-F slip catheter or angled J catheter is used for the occlusion with either the anchor or scaffold technique. Alternatively, if the right middle lobe or lingular PAVM has a small feeding artery, a standard microcatheter is placed through the 5-F JL 3.5 coronary catheter, and microcoils are used to close the PAVM.

Some interventionists prefer deploying a detachable fibered coil as the first coil and, now that the anchor tech-nique has been proven to be effective, we think there is little need for this to prevent paradoxical device embolization. Others have considered PAVMs as being similar to cerebral aneurysms and have preferred to occlude the aneurysmal sac rather than the feeding artery with expensive nonfibered detachable coils. Again, the physiology, anatomy, and considerations of preserving the parent artery are not present in PAVMs, as they are in cerebral aneurysms, and there is little indication for this approach.[12] Finally, the recent introduction of the Amplatzer vascular plug (both type 1 and 2) offers a single-device approach for closing PAVMs.[12] Although we have no experience utilizing the Amplatzer vascular plug, we follow its progress with interest. There may well be applications for its use for closing very large fistulae but, at the present time, teaching our fellows and junior staff the coaxial approach and the Anchor and Scaffold techniques has appeal because these techniques are widely applicable in all circulations in which large-vessel occlusion is required.

CONCLUSIONS

PAVMs become symptomatic in 50% of patients and can produce devastating neurologic complications. They should

be considered every time a patient with an ischemic embolic stroke or brain abscess presents to an emergency room. After excluding brain hemorrhage with an unenhanced CT of the brain, and if the CE yields positive results, PAVM as well as PFO should be in the differential diagnosis.

Because patients with PAVM usually have HHT, they should be referred after the acute problem for genetic counseling and possible treatment at one of the centers with specialized teams that manage this disorder on a daily basis. Alternatively, in an emergency situation, the PAVM can be closed effectively by application of precise and cost-effective means using coaxial catheters and the Anchor and Scaffold techniques with pushable fibered coils.

Results from HHT centers[5-10] suggest that these techniques are effective, and they are the basis for managing other patients requiring large-vessel occlusion. Treating the PAVM is just the tip of the iceberg for a family with an autosomal dominant disorder who needs screening and treatment. ■

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