2016

# Treating Pulmonary Arteriovenous Malformations: What to Do and What Not to Do

Ripal Gandhi
*Miami Cardiac & Vascular Institute*, gandhi@baptisthealth.net



Citation
Endovascular Today (2016) 15(4):52-55

# Treating Pulmonary Arteriovenous Malformations

What to do and what not to do.

**BY JUSTIN McWILLIAMS, MD, AND RIPAL T. GANDHI, MD**

Pulmonary arteriovenous malformations (AVMs) represent direct connections between the pulmonary artery and vein. Although they can be sporadic, approximately 70% of pulmonary AVMs are associated with hereditary hemorrhagic telangiectasia (HHT).[1] If left untreated, patients can present with paradoxical embolization (stroke or brain abscess), dyspnea and exercise intolerance, or pulmonary hemorrhage.[2] Embolization has become the favored method of treatment. This article serves to provide recommendations of what and what not to do in patients with pulmonary AVMs, based partially on published literature and partially on our personal experience in treating a multitude of these technically challenging lesions.

## WHAT TO DO

### Screen for HHT

Any patient with a pulmonary AVM has a high likelihood of HHT, and the presence of multiple pulmonary AVMs is almost pathognomonic of HHT. Ask any patient with pulmonary AVM the following two questions and perform the following examination:

- Do you have nosebleeds? More than 95% of adult patients with HHT have spontaneous and recurrent nosebleeds (median age of onset, 12 years).
- Do any of your family members get frequent nosebleeds? Because HHT is autosomal dominant (it does not skip generations), either the patient's mother or father is usually affected, and often other family members are as well.

Examine the lips, tongue, oral mucosa, and fingertips for small red spots. Telangiectasias are present in > 90% of adult patients with HHT, usually beginning in their 20s and increasing with age.

Any patient with a pulmonary AVM has a high likelihood of HHT, and the presence of multiple pulmonary AVMs is almost pathognomonic of HHT.

If any one of the three examination findings is positive, HHT is possible/probable, and if any two or three are positive, the diagnosis of HHT is definite, given that they already have a pulmonary AVM. In this case, referral to the closest HHT Center of Excellence (there are 22 in North America and several in Europe) should be considered.

### Think Before You Treat

Historically, a feeding artery diameter of 3 mm was considered to trigger embolization therapy for a pulmonary AVM.[2] Due to advancements in techniques and catheter technology, we now have the capability to treat lesions with feeding arteries as small as 1 mm. However, not all of these small AVMs require treatment. Complications of pulmonary AVMs < 2 mm are almost unheard of, and complications from AVMs in the 2- to 3-mm size range are uncommon.[3] Treating asymptomatic AVMs with a feeding artery of at least 3 mm is widely supported, as well as any symptomatic AVM. Asymptomatic AVMs under 2 mm are considered very low risk and are not typically treated. For asymptomatic 2- to 3-mm AVMs, embolization is generally recommended, but we often defer treatment in children and adolescents (with the plan to embolize at age 18), and in elderly patients.

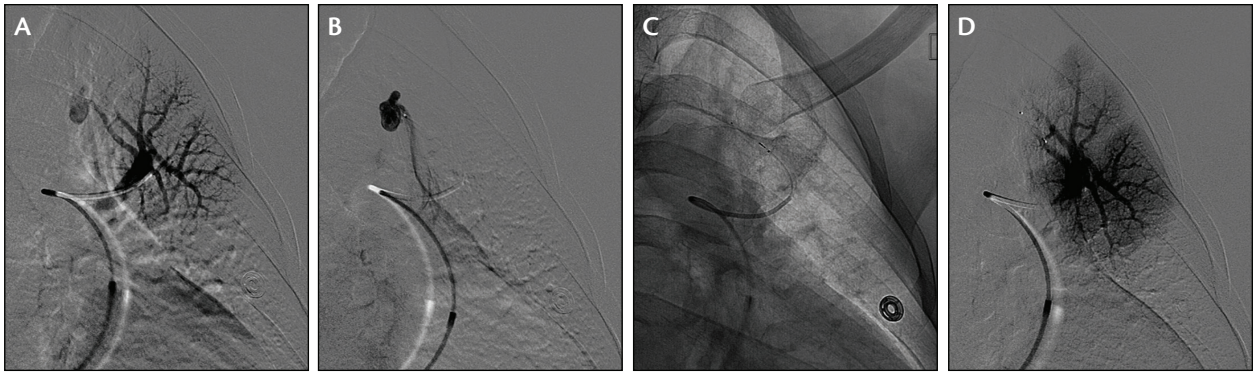


**Figure 1. This figure shows a 45-year-old man with HHT and asymptomatic 3-mm left upper lobe pulmonary AVM detected on routine screening. A left upper lobe angiogram demonstrates the AVM (A). Access to the left upper lobe artery was difficult, requiring a Judkins Right-4–shaped catheter. An angiogram through a 0.021-inch microcatheter demonstrates appropriate position at the proximal margin of the aneurysmal sac (B). Spot film demonstrating appropriate position of a MVP-3Q, prior to deployment (C). Final angiography confirms complete occlusion of the AVM with a single device (D).**

### Finding the Treasure Is Easiest With a Map

The procedure should commence with a high-volume angiographic run from the right or left pulmonary artery through a pigtail catheter. The ideal angle is determined by review of the preprocedure CT scan; if in doubt, a 30° to 40° contralateral oblique view opens and separates the pulmonary arteries most effectively. The best sensitivity for detecting pulmonary AVMs will come with imaging in full inspiration; however, this run will be almost useless as a "smart mask" for actual guidance into the target vessel. For intraprocedural guidance into the AVM itself, it is often more useful to ask the patient to "stop breathing" rather than to "take a breath in."

### Have the Right Stuff

Pulmonary AVM embolization is technically challenging because the constant movement of the heart and lungs tends to dislodge carefully placed catheters; wires tend to select the branch you do not want; and fear (or anxiety) is common at the moment of coil or plug release. For stability, we use a White Lumax catheter set (Cook Medical), which consists of a long, 7-F angled guide catheter to maintain stability in the parent artery and an inner 5-F angiographic catheter for vessel selection. If the origin of the desired vessel is particularly tortuous or difficult, a 5-F angled Glidecath (Terumo Interventional Systems) can be helpful. A Judkins Right-4 catheter (Merit Medical Systems, Inc.) is particularly effective for acute angled origins of the upper or middle lobes (Figure 1). It is preferable to navigate the angiographic catheter into the desired vessel using careful rotation and advancement of the catheter itself, checking the course with puffs of contrast. If this fails, an angled Glidewire (Terumo Interventional Systems) can be used to select the desired vessel, being careful not to push too aggressively.

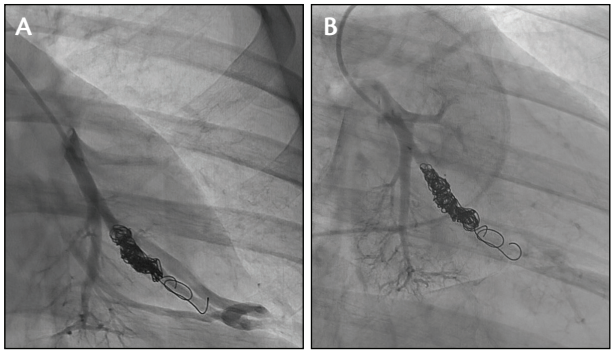


**Figure 2. This figure shows a 39-year-old man with HHT, after coil embolization of left lower lobe pulmonary AVM 5 years earlier. Initial angiography demonstrates recanalization through the previously placed coils (A). After deployment of a single MVP-5Q, the recanalized vessel is completely occluded (B). The adjacent pulmonary artery branches are spared.**

### Get Yourself Out There

Due to potential collateralization, AVM recanalization is common if embolization is performed too proximally. Distal embolization, either at the very distal aspect of the feeding artery[4] or within the AVM itself[5] is desired to minimize recurrence of the lesion. In AVMs with a large feeding artery, this can often be accomplished using Amplatzer Vascular Plugs (St. Jude Medical, Inc.) delivered through the 7-F guide catheter. These are typically oversized by 30% to 50%, with respect to the target vessel diameter. For intermediate-size AVMs, 0.035-inch coils can be delivered

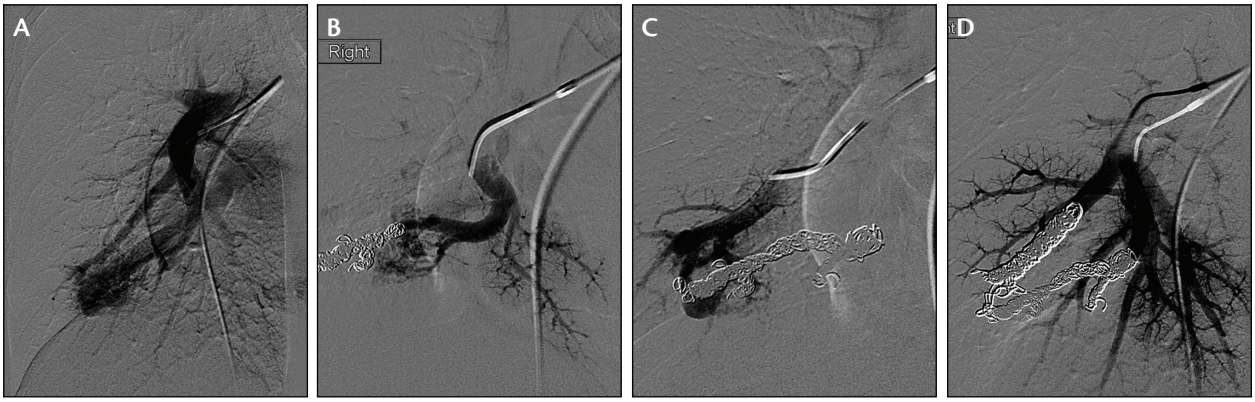


**Figure 3. This figure shows a 32-year-old woman with HHT and history of brain abscess. Initial pulmonary angiography confirms a complex AVM in the right lower lobe (A). Proximal embolization was considered to be high risk for recanalization. Control angiography after coiling of the distal portion of the anterior basilar segmental artery demonstrates persistent supply from the proximal portion (B). After coiling of this branch, angiography of the lateral basilar segmental artery demonstrates further supply to the AVM (C). Final angiography demonstrates complete occlusion of the AVM from its distal to proximal extent (D). A total of 36 coils were used.**

through a 4- or 5-F catheter, an Amplatzer Vascular Plug 4 can be delivered through a 5-F angiographic catheter, and MVP-7Q and MVP-9Q microvascular plugs (Medtronic) can be delivered through 4- and 5-F catheters, respectively. For small or tortuous AVMs, distal embolization usually requires a microcatheter. It is important to use a microcatheter size that is compatible with your desired embolic agent (a standard microcatheter for standard microcoils or a high-flow microcatheter for larger microcoils). Try to avoid smaller microcatheters, which provide suboptimal angiographic runs and can be easily misdirected into tiny pulmonary sub-branches.

We have found the MVP-3Q and MVP-5Q to be very useful for single-device occlusion of small AVMs. The MVP-3Q occludes vessels up to 3 mm and is compatible with standard-size microcatheters, whereas the MVP-5Q occludes vessels up to 5 mm and is compatible with high-flow microcatheters. For complex AVMs, long detachable coils can be used to embolize the AVM nidus to prevent recanalization (Figure 2). We have found that the standard and soft Interlock fibered coils (Boston Scientific Corporation) provide a good balance of rapid deployment, efficient occlusion, and relatively low cost. Concerto coils (Medtronic) are very soft, reducing pushback and making them useful when the microcatheter is tenuously positioned. Other good detachable coil alternatives include Ruby coils (Penumbra, Inc.) and Azur hydrocoils (Terumo Interventional Systems). For pushable coils, the Nester and MicroNester line of coils (Cook Medical) provide inexpensive cross-sectional occlusion.

### Beware the Air (and the Clot)

An air embolism is a serious risk when treating pulmonary AVMs; any air passing into a pulmonary AVM can pass directly into the left-sided circulation and from there, into the brain, causing a transient ischemic attack or worse. Prior to the procedure, nurses should be cautioned not to inject any air bubbles when the IV is started, and an air filter should be placed if available. During the procedure, blood should be aspirated prior to each injection, even with the microcatheter, to ensure a solid liquid column within the catheter. Wires should be removed either under saline within a basin or with a constant drip of saline into the catheter hub. Although there is institutional variation, we choose to administer heparin to all patients immediately after venous access to prevent clot formation on the catheter during treatment.

## WHAT NOT TO DO

### Don't Test Your Luck

The moment of delivery of the first coil is often one of consternation. To minimize the stress, use a long, detachable, and slightly oversized coil for the first coil, and watch that it deforms as you push it out (suggesting ample wall contact) and that it is completely immobile prior to full deployment (suggesting secure position). If there is any concern, simply recapture the coil and choose a more appropriate size. Pushable coils can be used behind the first coil, if desired, but the risk/reward ratio greatly favors that the all-important first coil be detachable. Alternatively, use a vascular plug, all of which are detachable and secure if sized properly (if you are unsure, use a larger size).

### Don't Skimp on the Embolic Agent

Recanalization occurs either by collateralization around the embolic agent, or by flow resumption directly through the embolic agent. To minimize the latter, create a tight coil pack composed of multiple coils, and if in doubt, add more. If a porous plug, such as an Amplatzer Vascular Plug, is used, backing it up with one or two cheap pushable coils will help prevent recanalization through the plug. Polytetrafluoroethylene-lined plugs, such as the MVP, can be a one-device solution (Figure 3), but may not produce full occlusion if placed in areas of tortuosity and can also be backed up with coils in this instance.

### Don't Sacrifice Safety for Convenience

Being sure not to sacrifice safety is especially important when treating multiple lesions in one session because radiation and contrast limits may come into play. There is no shame in staging the procedure and bringing the patient back on a different occasion to finish the job.

## CONCLUSION

Treatment of pulmonary AVMs is challenging and exhilarating. Proper planning and appropriate equipment, along with good working knowledge of what (and what not) to do, will help optimize outcomes. ■


1. Gossage JR, Kanj G. Pulmonary arteriovenous malformations: a state of the art review. Am J Respir Crit Care Med. 1998;158:643-661.
2. White RI Jr, Pollak JS, Wirth JA. Pulmonary arteriovenous malformations: diagnosis and transcatheter embolotherapy. J Vasc Interv Radiol. 1996;7:787-804.
3. Trerotola SO, Pyeritz RE. PAVM embolization: an update. Am J Roentgenol. 2010;195:837-845.
4. Pollak JS, Saluja S, Thabet A, et al. Clinical and anatomic outcomes after embolotherapy of pulmonary arteriovenous malformations. J Vasc Interv Radiol. 2006;17:35-44.
5. Kajiwara K, Urashima M, Yamagami T, et al. Venous sac embolization of pulmonary arteriovenous malformation: safety and effectiveness at mid-term follow-up. Acta Radiol. 2014;55:1093-1098.



**Justin McWilliams, MD**
David Geffen School of Medicine
University of California
Los Angeles, California
(310) 267-8773; jumcwilliams@mednet.ucla.edu
*Disclosures: None.*

**Ripal T. Gandhi, MD**
Miami Cardiac and Vascular Institute
Baptist Health South Florida
Miami, Florida
*Disclosures: Consultant for Medtronic.*