

# Management of Intraosseous AVMs

Because bone vascular AVMs are rarely seen in day-to-day practice, clinicians should be aware of the steps for proper diagnosis and the available treatment options to improve patient outcomes.

BY WAYNE F. YAKES, MD

Vascular malformations constitute some of the most difficult diagnostic and therapeutic dilemmas that can be encountered in the practice of medicine. The clinical presentations are protean and can range from an asymptomatic birthmark to fulminant life-threatening congestive heart failure. Attributing any of these varied symptoms that a patient may present with to a vascular malformation can be challenging to the most experienced clinician. Compounding this problem is the rarity of these vascular lesions. If a clinician sees one patient with a vascular malformation every few years, it is difficult to gain a learning curve to diagnose and optimally treat him or her. Typically, these patients bounce from clinician to clinician only to experience disappointing outcomes, complications, and recurrence or worsening of their presenting symptoms.

Surgeons initially treated vascular malformations. The early rationale of proximal arterial ligation of arteriovenous malformations (AVMs) proved futile as the phenomenon of neovascular recruitment reconstituted arterial inflow to the AVM nidus; microfistulous connections became macrofistulous feeders. Complete extirpation of a vascular malformation can be difficult and, at times, even hazardous, necessitating suboptimal partial resections. Partial resections can cause an initial good clinical response and may last for some time. However, often the patient's presenting symptoms recur or worsen at follow-up.<sup>1-3</sup> Because of the significant blood loss that frequently accompanies surgery, the skills of interventional radiologists were eventually employed to embolize these vascular lesions preoperatively. This allowed much less blood loss and a more complete resection to be performed. However, complete extirpation of a vascular malformation is still a difficult issue and may not always be possible. Various authors have published that only approximately 20% of malformations may be amenable to complete extirpation with surgery.<sup>2,4,5</sup>

According to D. Emerick Szilagyi, MD, former editor for the *Journal of Vascular Surgery*, "...with few exceptions, their (vascular anomalies) cure by surgical means is impossible. We intuitively thought that the only answer of a surgeon to the problem of disfiguring, often noisome, and occasionally disabling blemishes and masses, prone to cause bleeding, pain, or other unpleasantness, was to attack them with vigor and with the determination of eradicating them. The results of this attempt of radical treatment were disappointing."<sup>2</sup> Indeed, of the 82 patients seen in this patient series, only 18 patients were deemed operable, and of the 18 operated upon, 10 were improved, two were the same, and six were worse at follow-up.

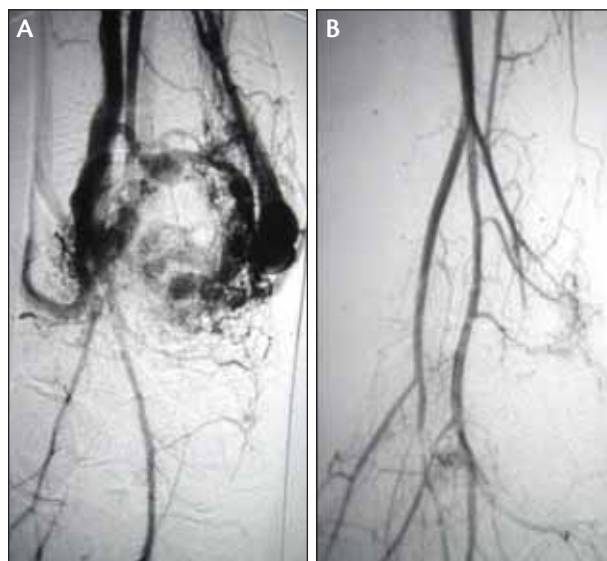
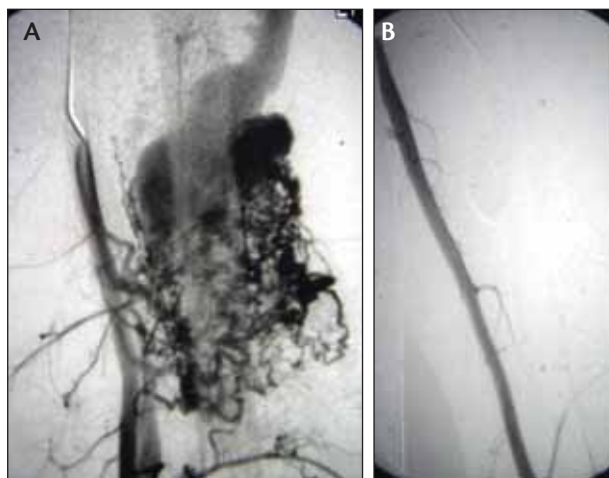


Figure 1. A right popliteal arteriogram showing an intraosseous AVM involving the talus and calcaneus bones (A). A right popliteal arteriogram 3 years after ethanol embolization, showing the absence of the AVM involving the talus and calcaneus bones (B).



**Figure 2.** A right femoral arteriogram showing a massive intraosseous AVM of the right femur (A). A right femoral arteriogram 6 years after ethanol and coil embolization showing the absence of the massive femur AVM (B).

### CONCEPTS IN PATIENT MANAGEMENT

Vascular malformations are congenital lesions that are present at birth, whether evident clinically, and grow commensurately with the child. The collective term *vascular malformation* encompasses any malformed blood vessel of any vascular element to include arteries, capillaries, veins, and lymphatics. Posttraumatic arteriovenous fistulae (AVF) are different in that they are an acquired lesion, although they may be similar radiographically.<sup>6,7</sup> A thorough clinical exam and history can usually establish the diagnosis of hemangioma or vascular malformation.

Color Doppler imaging (CDI) is an essential tool in the diagnostic workup of vascular malformations. Both high-flow lesions (AVMs, AVFs) and low-flow lesions (venous malformations, lymphatic malformations) can be accurately diagnosed. Furthermore, CDI is an important noninvasive method for following patients who have undergone therapy. Documentation of increased arterial flow rates before therapy and decreased arterial flow rates after therapy provides important physiological information. In low-flow malformations, persistent documentation of thrombosis can be imaged and accurately assessed.<sup>8</sup>

Computed tomography (CT), although helpful in the diagnostic workup, is less useful than magnetic resonance imaging (MRI). Unlike CT, MRI easily distinguishes between high-flow and low-flow vascular malformations. Furthermore, the anatomic relationships of the vascular malformation to adjacent nerves, muscles, tendons, organs, bone, and subcutaneous fat allow a total assessment. MRI is also an excellent noninvasive method for following patients to determine the efficacy of therapy, often obviating repetitive arteriography and venography.<sup>9</sup> CT has its main role in

intraosseous vascular malformations and determination of the cortical margins and its involvement (Figures 1 and 2).

With the use of ethanol as an embolic agent to treat vascular malformations in all anatomic locations, patients require general anesthesia. At times, Swan-Ganz line and arterial-line monitoring may be necessary when treating larger lesions with large amounts of ethanol. Pulmonary artery pressures then can be constantly monitored to determine if abnormal elevation of the pulmonary pressures occurs during the procedure. The embolization procedure can be stopped at that point to allow the pulmonary pressures to normalize. If the pressures are high enough, the infusion of nitroglycerine through the Swan-Ganz catheter can help lower the pulmonary artery pressures to normalcy. By injecting 0.1 mL of ethanol every 10 minutes, using the Swan-Ganz lines is rarely necessary.

### ENDOVASCULAR THERAPY OF BONE VASCULAR MALFORMATIONS

Embolization procedures have evolved as one of the cornerstones of modern interventional radiology. Now that it is firmly established as an essential therapeutic tool, its role will only continue to grow. There are many embolic agents that are used in various clinical scenarios. With regard to vascular malformations, permanence is a significant issue. It has already been documented in the literature that embolization with polyvinyl alcohol, tissue adhesives, coils, Onyx (ev3 Inc., Plymouth, MN), and the like are rarely curative and will only provide palliation.<sup>10-12</sup> With the advent of the use of ethanol, cures at long-term follow-up have been documented by multiple authors.<sup>13,14-25</sup> The judicious use of ethanol as an embolic agent has revolutionized our abilities to permanently cure these lesions in the soft tissues, organs, bone, and brain.<sup>13,14-25</sup>

AVMs of soft tissues and bones pose a significantly difficult dilemma. They are typified by hypertrophied in-flow arteries shunting through a primitive vascular nidus into tortuous dilated outflow veins. No intervening capillary bed is present. Symptoms are usually referable to the anatomic location of the AVM. The larger and more anatomically central an AVM is, the greater the likelihood of high-output cardiac consequences. Other presenting symptoms can include pain, progressive nerve deterioration or palsy, disfiguring mass, tissue ulceration, hemorrhage, impairment of limb function, limiting claudication, and so on.

Endovascular therapy of AVMs with ethanol has ushered in a new era in the therapy of these problematic congenital anomalies. Cures and permanent partial ablations have been documented in our patient series resulting in dramatic symptomatic improvement.<sup>13,14-25</sup> Because neovascular recruitment and recanalizations have not been observed, permanent partial ablations have also led to long-term symptomatic improvement, obviating the need

for further treatment. Despite the success that is possible with ethanol, it must be remembered that it is an extremely dangerous intravascular sclerosant that can cause total tissue devitalization and neuropathy.<sup>26-32</sup>

Intraosseous AVMs can be extremely challenging because of their position within bone. At times, transarterial routes may suffice to gain access into the bone AVM itself; however, this is not always possible. Frequently, the inflow artery turns into a meshwork rather than a direct tube into the AVM obviating a transcatheter approach. At this point, where CT could be helpful in defining the thinnest point of bone, direct puncture can be performed with a needle to access the malformation within bone, and ethanol can then be utilized.

Other techniques for gaining access to the AVM nidus within bone may be through a transvenous approach where the vein exits the bone; by taking advantage of that, the interventionist can access via the retrograde position from the vein to the area of fistulization in the vein within the bone AVM. Tourniquets, blood pressure cuffs, or occlusion balloons may be helpful in slowing down the vascular flow to allow greater contact time of the ethanol within the malformation to denude the endothelium, fracture the blood vessel wall to the level of the internal elastic lamina, and thus cause platelet aggregation and thrombosis of the vessel from the wall that is denuded centrally. Despite inflow occlusion, the flows may still be so great that other techniques will be necessary. One such technique is gaining access of the vein area of the fistulization, placing a few fiber coils to slow down the flow, and then injecting the ethanol and ultimately occluding it with coils. This is also a helpful technique in these high-flow lesions.

Complications are part and parcel with treating vascular malformations, whether surgically or endovascularly. It is crucial for a vascular malformation team to be in place so that complications, when they occur, can have their morbidity dramatically reduced. At my institution from January 2002 to December 2006, I performed 5,889 procedures on 1,247 patients using 111,742 mL of ethanol. The transient complication rate was 5%, and the major complication rate was 1%.

## CONCLUSION

AVMs pose a significant challenge in the practice of medicine today. Bone AVMs cause unique clinical problems with regard to their anatomic location. Clinical manifestations of these lesions are extremely protean. Because of the rarity of these lesions, the experience of most clinicians is limited, and therefore so is the diagnosis and management, often leading to misdiagnoses and poor patient outcomes, augmenting the enormity of the problem. AVMs are best treated in medical centers where patients with these lesions are seen regularly and the team approach is used. Only in this fashion can significant experience and improvement in

judgment in managing these lesions develop definitive statements for the treatments of AVMs. Endovascular management of bone AVM is a limb-sparing procedure. ■

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