Arteriovenous Malformations: Definitions, Presentation, Imaging, and the Role of Embolization

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Introduction:

Vascular malformations (VM) are benign defects of vascular morphogenesis which can be divided by low flow and high flow characteristics. Arteriovenous malformations (AVM) are a high flow VM that develop during embryogenesis or acquired at any stage after birth.

Little is known about preventing development of AVMs, though timely treatment prevents AVM progression and deleterious outcomes. Left untreated, AVMs may progressively grow. The continuous arteriovenous shunting may progress to high-output heart failure.

Characterization:

AVM classification may be based on presenting symptoms (Table 1) or by response to endovascular treatment strategies (Table 2).

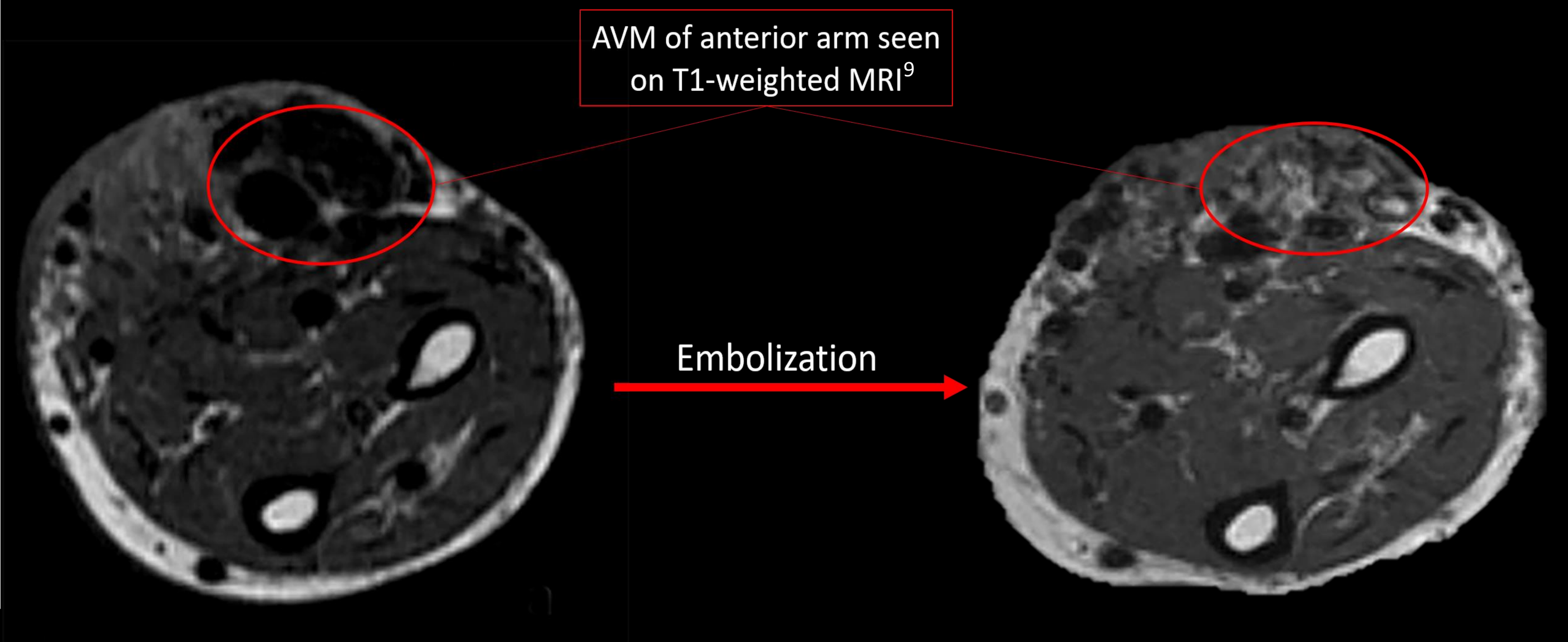

Table 1: Schobinger Classification[1]

Type 1	Quiescent: Warm, pink-blue, shunting on Doppler
Type 2	Expansion: Enlargement, pulsation, thrill, bruit, tortuous veins
Type 3	Destruction: Dystrophic skin changes, ulceration, bleeding, pain
Type 4	Decompensation: High-output cardiac failure

Table 2: Cho Classification[2]

Type I	Arteriovenous fistulae → best responds to retrograde or direct puncture
Type II	Arteriolovenous fistulae →best responds to retrograde or direct puncture
Type IIIa	Arteriolovenulous fistulae with non-dilated fistula → trans-arterial or direct puncture approach preferred
Type IIIb	Arteriolovenulous fistulae with dilated fistula → trans-arterial or direct puncture approach preferred

Role of Imaging in AVM Diagnosis

Initial imaging tests are often CT with contrast, MRI with contrast, or Doppler ultrasound MRI/MRA are often preferred primary imaging modalities for its anatomical resolution of AVM and surrounding soft tissue	
Doppler US[3]	Arterial waveforms High flow venous structures (indicates vascular shunting)
CT[4]	CTA is alternative to MRA 4D-CTA assists with three-dimensional reconstruction
MRI[5]	T1- and T2- weighted images show conglomerate of flow voids Time-resolved MRA helps to delineate nidal anatomy and assess treatment efficacy
Angiography[6]	Should be performed on almost all AVMs Assess flow rate, visualizes anatomy of nidus, and identifies vessels required for distal circulation Predicts treatment response based on nidal architecture

US = ultrasound; CT = computed tomography, CTA = CT angiography; MRI = magnetic resonance imaging; MRA = MR angiography

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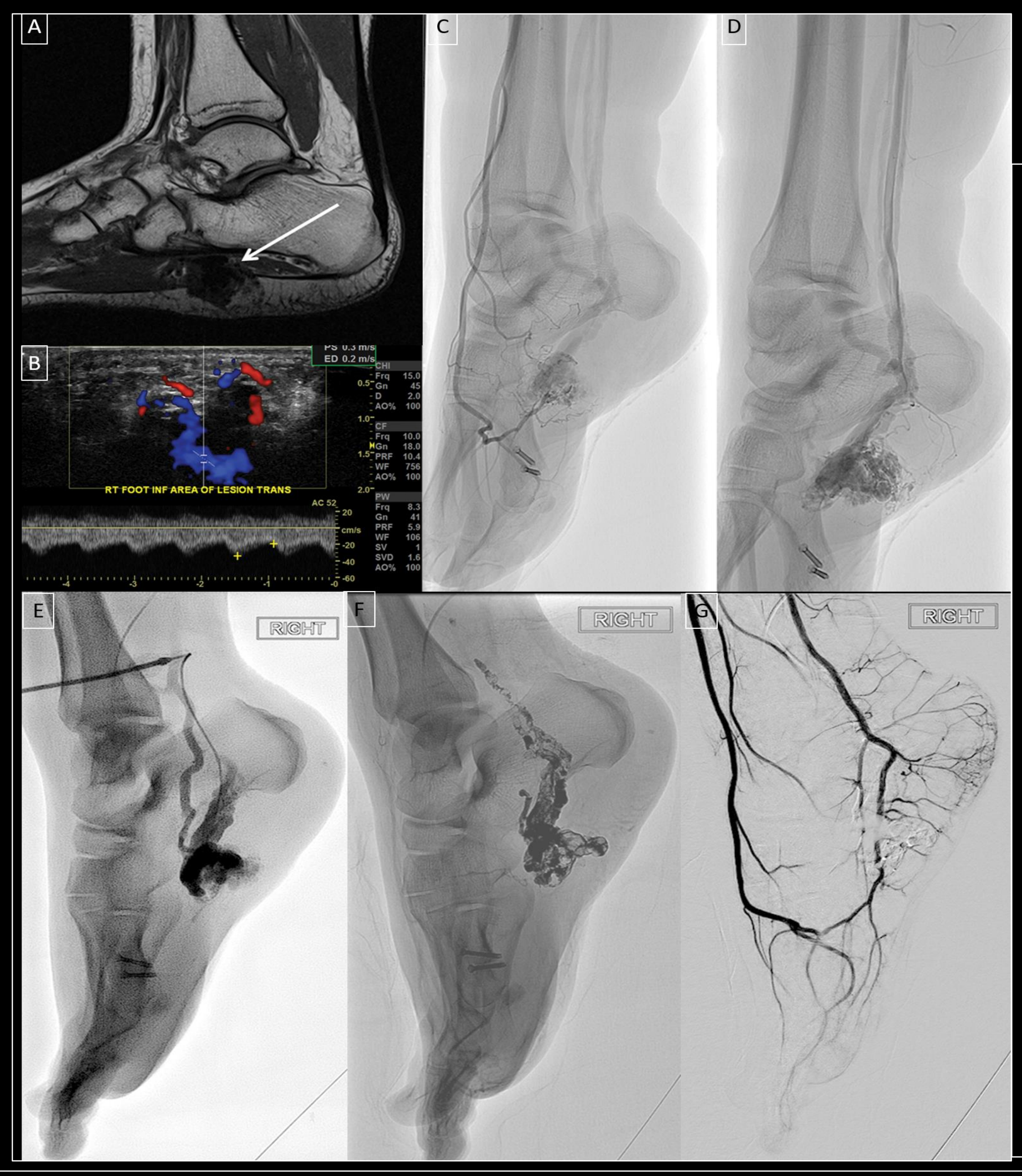

Figure 2
A: Sagittal T1-weighted MRI showing flow voids on sole of foot
B: Doppler ultrasound showing arterial waveforms
C/D: Selective angiogram of anterior (C) and posterior (D) tibial artery
E: Diagnostic venogram showing contrast stasis in AVM
F: Embolization with NBCA
G: Postembolization DSA with complete occlusion of AVM

Modified from Funaki B and Funaki C (2016)[10]

Endovascular Treatment (Embolization)

Oftentimes, multiple different embolics are used to treat single AVM. Embolic materials include:

Ethanol[7,8]
- Widely used, but may need to be diluted or avoided in AVMs involving large portions of skin to reduce risk of edema, skin necrosis, and nerve damage
- Dose-dependent risk of pulmonary hypertension and cardiovascular collapse → monitor pulmonary arterial pressure
- Procedure is very painful → general anesthesia is required

N-butyl cyanoacrylate (NBCA)[9]
- Liquid casting adhesive agent that quickly polymerizes and irreversibly when exposed to anions
- Preferred in AVMs with large, draining veins (that would require great amounts of ethanol or Onyx)
- Preferred in pediatric patients

Poly-vinyl-alcohol (PVA) particles[9]
- Used as pre-surgical adjunct or management of acutely bleeding AVM

Ethylene vinyl alcohol copolymer (Onyx)[9]
- Liquid casting adhesive agent for central nervous system AVMs
- Primary advantage over NBCA is slower flow and longer casting time

Endovascular coils and vascular plugs[9]
- Limited role in peripheral AVMs due to their size which may limit future vascular access if subsequent embolization is required

Treatment Techniques

Goal of AVM embolization = obliterate nidus but reduce off-target embolization
- Best achieved by slowing flow → improves operator control and assists with catheter positioning

Treatment approach based on angiographic findings (Table 2)[2,9]
- AVM with dominant outflow vein (types 1 and 2) → retrograde or direct puncture preferred
- Large aneurysmal draining veins → coils and glues preferred
- Small AVMs → NBCA potentially curative
- AVMs with multiple feeders and outflows (types 3a and 3b) → trans-arterial or direct puncture approach preferred