INTRODUCTION

Arteriovenous malformations (AVMs) are congenital vascular malformations (CVMs) caused by birth defects involving the arterial and venous origins that result in direct communications between vessels of different sizes or primitive reticular networks of dysplastic minute vessels that have failed to mature into capillary vessels. Based on angiographic findings, peripheral AVMs can be classified into six types: type I, type IIa, type IIb, type IIc, type IIIa, and type IIIb. Treatment strategies vary with the types. Type I is treated by embolizing the fistula between the artery and the vein with coils. Type II (IIa, IIb, and IIc) AVM is treated as follows: first, reduce the blood flow velocity in the venous segment of the AVM with coils; second, perform ethanol embolotherapy of the residual shunts. Type IIIa is treated by transarterial catheterization of the feeding arteries and injection of diluted ethanol. Type IIIb is treated by transarterial or direct puncture approaches. A high concentration of ethanol is injected through the transarterial catheter or direct puncture needle. When the fistula is large, coil insertion is required to reduce the amount of ethanol. Type I and type II AVMs showed the best clinical results; type IIIb showed a satisfactory response rate. However, type IIIa showed the poorest response rate, either alone or in combination with other types. Clinical success can be achieved by using different treatment strategies for different angiographic AVM types.

Keywords: Arteriovenous malformation; Embolotherapy; Angiography

How to Treat Peripheral Arteriovenous Malformations

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Arteriovenous malformations (AVMs) are direct communications between primitive reticular networks of dysplastic vessels that have failed to mature into capillary vessels. Based on angiographic findings, peripheral AVMs can be classified into six types: type I, type IIa, type IIb, type IIc, type IIIa, and type IIIb. Treatment strategies vary with the types. Type I is treated by embolizing the fistula between the artery and the vein with coils. Type II (IIa, IIb, and IIc) AVM is treated as follows: first, reduce the blood flow velocity in the venous segment of the AVM with coils; second, perform ethanol embolotherapy of the residual shunts. Type IIIa is treated by transarterial catheterization of the feeding arteries and injection of diluted ethanol. Type IIIb is treated by transarterial or direct puncture approaches. A high concentration of ethanol is injected through the transarterial catheter or direct puncture needle. When the fistula is large, coil insertion is required to reduce the amount of ethanol. Type I and type II AVMs showed the best clinical results; type IIIb showed a satisfactory response rate. However, type IIIa showed the poorest response rate, either alone or in combination with other types. Clinical success can be achieved by using different treatment strategies for different angiographic AVM types.

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and outcomes (4, 7-9). In this article, the angiographic classification and endovascular treatment of peripheral AVMs will be described.

**Angiographic Classification of Peripheral AVMs**

Angiography is the gold standard for confirmative diagnosis of AVM as well as therapeutic guidance. Typically, angiography of AVMs shows feeding arteries, nidus, and early draining veins. Angiographic findings of AVMs differ in every patient; the malformed vasculature is very complex, and it is difficult to figure out the detailed vascular connection at a glance. Therefore, the classification of AVMs has been necessitated to guide treatment and evaluate outcomes. The simplified angiographic classification of intracranial AVMs, proposed by Houdart et al. (10), is considered adequate for application to peripheral AVMs. In this simplified classification system, AVMs were classified into three mainly based on the number and sizes of the feeding arteries and veins as well as the vascular connections between them. Based on this concept, Cho et al. (4) and Ko et al. (8) proposed a modified angiographic classification for AVMs in the torso and the extremities in 2006 and 2019, respectively.

Figure 1A describes the vascular anatomical connections between the feeding arteries and draining veins. Type I AVMs are characterized by arteriovenous fistulae that consist with three or less feeding arterial shunts to a single draining vein that is a single direct arteriovenous fistula. Type II AVMs are characterized by arteriovenous fistulae that consist with multiple arteriolar shunts to the single draining vein. According to the morphology of the draining vein, type II is subclassified into types IIa, IIb, and IIc (8). Type IIa AVMs are characterized by multiple arteriolar shunts to the focal segment of the single draining vein. Type IIb AVMs are characterized by multiple arteriolar shunts to the venous sac with multiple draining veins. Type IIc AVMs are characterized by multiple arteriolar shunts along the long segment of the draining vein. Type III AVMs have multiple shunts between the arterioles and venules; when the fistula of the nidus is observed as a blush or fine striaion on angiography, it is categorized as type IIIa for

![Figure 1A](https://example.com/fig1a.png)

**Fig. 1. Angiographic classification and treatment strategy of peripheral AVMs.**

A. Angiographic classification of AVMs. Type I: arteriovenous fistulae with three or less feeding arteries and a single draining vein. Type IIa: multiple arterioles shunt to the focal segment of the single draining vein. Type IIb: multiple arterioles shunt to the venous sac with multiple draining veins. Type IIc: multiple arterioles shunt along the long segment of the draining vein. Type IIIa: multiple arterioles shunt to multiple draining veins through multiple fine fistulae. Type IIIb: multiple arterioles shunt to multiple draining veins through multiple enlarged fistulae.

B. Strategies for embolotherapy based on the types of AVMs. Type I: arteriovenous fistula is embolized with coils. Type IIa: the focal venous sac is embolized with coils or core-removed guidewires through the direct puncture or a transvenous catheter approach. Type IIb: the venous sac is embolized with coils or core-removed guidewires through the direct puncture approach. Type IIc: the long segment of the draining vein is occluded with coils or core-removed guidewires using the transvenous approach or a direct puncture. Type IIIa: diluted ethanol (50–60%) is injected through the transarterial catheter. Type IIIb: a high concentration of ethanol (80–100%) is injected through the transarterial catheter or the direct puncture needle. When the fistula is large, coil insertion is required to reduce the amount of ethanol. A = artery, AVM = arteriovenous malformation, DP = direct puncture, S = shunt, TA = transarterial, TV = transvenous, V = vein
a non-dilated fistula, and when the fistula of the nidus is observed as a complex vascular network, it is classified as type IIIb for a dilated fistula (Figs. 2-7).

**Endovascular Treatment**

Based on the angiographic classification of AVMs, the treatment strategies should vary. Figure 1B shows the typical treatment approach and embolotherapy based on the angiographic types of AVMs. The main target of type I AVMs is the direct fistula between the artery and the draining vein. The fistula is embolized with coils or plugs by the transarterial approach (Fig. 2). Generally, our treatment strategy for type II AVMs is as follows: first, reduce the blood flow velocity in the venous segment of the AVM with coils; second, perform ethanol embolotherapy of the residual shunts, because the shunts are located at the venous wall of the draining vein. For a type IIa AVM, the focal venous segment of the draining vein of the AVM is approached using a direct puncture or transvenously and embolized by packing with coils or a core-removed guidewire. Because coil packing is not enough to occlude the shunts, an additional injection of high-concentration ethanol (80–100%) is required through a transarterial superselective microcatheter, percutaneous puncture needle, or transvenous catheter (Fig. 3). For a type IIb AVM, a direct puncture of the venous sac is considered the most effective approach. Coil packing of the venous sac followed by ethanol injection is similar to that of a type IIa AVM. When the venous sac is small, the ethanol injection is enough to occlude the AVM (Fig. 4). For a type IIc AVM, the long segment of the draining vein is occluded with coils or core-removed guidewires using the transvenous or direct puncture approach. Between the insertion of multiple coils, ethanol should be injected into the vein to occlude the shunts at the venous wall (Fig. 5). A type IIIa AVM is treated by transarterial catheterization of the feeding arteries and the injection of diluted ethanol (50–60%), which has been mixed with a contrast medium because micro fistulae are too small to puncture directly (Fig. 6). Most of the type IIIa AVMs are located within the superficial area (involvement of skin or subcutaneous tissue), and intra-arterial injection of a high concentration of ethanol results in serious skin necrosis. Therefore, we generally use low concentrations of ethanol (diluted with contrast medium to 50–60%) with caution, rather than pure ethanol or high concentrations of ethanol. If there is no therapeutic improvement after the injection of diluted ethanol during several sessions, we consider using compression stocking to relieve swelling and pain. A type IIIb AVM is treated using transarterial or direct puncture approaches. A high concentration of ethanol (80–100%) is injected through

**Fig. 2. Images of a 42-year-old woman with a type I renal AVM.**

A. Venous phase of the pretreatment angiogram shows the direct arteriovenous fistula formation at two intrarenal branches. B. A selective angiogram of the proximal portion of arteriovenous fistula using a microcatheter shows a type I AVM more clearly. Arteriovenous fistulae were embolized with coils using the intra-arterial approach. A total of 12 coils were inserted into the feeding arteries of individual arteriovenous fistulae. C. Completion angiogram shows complete obliteration of the AVM. Other normal intrarenal branches were completely spared without flow disturbance. The treatment was completed in a single session. The AVM did not recur during the 6-year follow-up.
the transarterial catheter or direct puncture needle. When the fistula is large, coil insertion is required to reduce the amount of ethanol (Fig. 7). Intra-arterial embolization is the preferred technique during the initial periods of type IIIb treatment. However, an intra-arterial ethanol injection is prone to creating skin complications because tiny angiographically invisible normal arteries become exposed to ethanol. Direct puncturing of the dilated fistula and injecting ethanol were proven to be effective, and they induced fewer skin complications (9).

Since the initiation of endovascular therapy for AVMs, ethanol has been the preferred embolic agent because of its strong devascularization effect resulting from endothelial damage of the vessel, serum protein denaturation, and
Fig. 4. Images of a 27-year-old man with a type IIb foot AVM.
A. The venous phase of a pretreatment angiogram shows multiple arterioles from the anterior and posterior tibial arteries shunting to the venous sac with multiple small draining veins. There are two small aneurysmal changes (asterisks) in the anterior tibial artery. B. A selective angiogram after the puncture of the venous sac shows the venous sac with multiple small draining veins more clearly. C. A total of 15 cc of pure ethanol was injected slowly, and a blood pressure cuff was inflated to 150 mm Hg. D. A completion angiogram shows complete obliteration of the AVM. The treatment was completed in a single session. The AVM did not recur during the 6-year follow-up.

Fig. 5. Images from a 6-year-old boy with type IIc AVM in the thigh.
Arterial (A) and venous (B) phases of a pretreatment superficial femoral artery angiogram show multiple arterioles shunting along the long segment of the draining vein (asterisk). The long segment of the draining vein was occluded with coils using the transvenous and direct puncture approach. After slowing the flow within the AVM, pure ethanol was injected using the transvenous and intra-arterial approaches to eradicate the residual AVM lesion (C). A completion angiogram of the common femoral artery shows complete obliteration of the AVM. The AVM did not recur during the 6-year follow-up (D).
rapid thrombus formation (2-4, 11). However, ethanol induces significant pain in the vessel wall vasa nervorum when injected intravascularly. General anesthesia is required to minimize patient discomfort. Post-embolization edema always occurs with the use of ethanol. Caution with superselective catheter/needle positioning should be exercised to minimize the possibility of non-target embolization of normal tissues to prevent tissue necrosis and neuropathy. Pulmonary hypertension is a potentially fatal complication associated with ethanol embolotherapy, and it occurs when a significant amount of ethanol is injected. The etiology of pulmonary hypertension is related to precapillary pulmonary artery spasm or extensive microthromboembolism. Pulmonary hypertension can lead to a cardiopulmonary arrest if it is not controlled effectively. To reduce the risk of cardiopulmonary complications during

Fig. 6. Images of a 16-year-old girl with a type IIIa AVM in the shoulder. A. Venous phase of a pretreatment angiogram shows the AVM of the nidus as a diffuse blush and early draining veins. B. Superselective angiogram shows microfistulae between the arterioles and venules as a blush. C. Completion angiogram (after five sessions of intra-arterial injection of 50–70% diluted ethanol through the fine feeding arteries) shows a markedly improved AVM.

Fig. 7. Images of a 43-year-old man with a type IIIb AVM in the foot. A. Venous phase of a pretreatment angiogram shows multiple dilated fistulae and dilated draining veins. B. Direct puncture of one of the dilated fistulae shows typical hypertrophy of the fistula, which was compatible with a type IIIb AVM. Pure ethanol was injected directly through the needle, and the blood pressure cuff was inflated to 150 mm Hg. C. Completion angiogram (after three sessions of an injection of 80–100% ethanol through the needle) shows near-complete embolization of the foot AVM with an intact dorsalis pedis branch (arrow).
ethanol embolotherapy, appropriate measures should be taken, which include the administration of general anesthesia and close cardiopulmonary monitoring. When a high volume of ethanol injection (more than 0.5 mL/kg) is planned, pulmonary artery Swan-Ganz line and arterial line monitoring are recommended to minimize the risk of this event. When the mean pulmonary artery pressure (PAP) is elevated by more than 25 mm Hg, the infusion of nitroglycerine is recommended as a bolus injection (50–100 μg) and as a continuous infusion (0.3–3.0 μg/kg/min) through the Swan-Ganz line (2-4, 12, 13). Our recent recommendations for ethanol injection are as follows: 1) a single bolus injection of ethanol is limited to 0.1 mL/kg of bodyweight, and the maximum volume of ethanol should be limited to 1.0 mL/kg of bodyweight; 2) after intravascular ethanol injection, a 10-minute waiting time until the next injection is recommended to observe the thrombotic effect and avoid the systemic effect of ethanol; 3) when the expected total volume of ethanol is less than 0.5 mL/kg of bodyweight, monitoring of PAP is not required.

Endovascular treatment of AVMs using other liquid embolic agents such as n-butyl cyanoacrylate (NBCA) and Onyx has been reported. The symptomatic improvement in NBCA is good, but the clinical result is not long-lasting (14). Onyx is a more controllable agent, but its reported data are limited, and it is associated with a risk of recanalization and recurrence. The drawbacks of Onyx are high costs, radioopacity, and residual mass effects (15, 16).

**Treatment Results**

Based on the reported data of Ko et al. (8) and Park et al. (9), the overall cure rate in 306 patients with peripheral AVMs was 39%, and the clinical success rate (cure + markedly improved) was 60%. Among them, type I AVMs were rare (less than 2%), but the cure rate after fistula embolization was 100%. The proportion of type II AVMs was 27%, and the cure rate was up to 80%; the cure rate of type IIa (95%) was significantly better than that of type IIc (65%; \(p = 0.015\)) AVMs. The proportions of types IIIa and IIIb were 5% and 40%, respectively. Types IIIa and IIIb showed less favorable therapeutic responses to embolotherapy. The cure rates of types IIIa and IIIb were 19% and 30%, respectively, and their clinical success rates were 50% and 56%, respectively. Although the angiographic classification system provided distinguishable categories for the AVMs, not all of them completely fall into the six specified types. Twenty-seven percent of AVMs are classified as complex, as they belong to different categories at the same time (9). Based on the angiographic classification, patients with type I and type II AVMs showed the best clinical results after ethanol embolotherapy with or without coils; those with type IIIb AVMs showed satisfactory outcomes. However, type IIIa AVMs showed the poorest response, either alone or in combination with other types. One study showed that the extent of a lesion is associated with treatment response (7). Even with type IIIb AVMs, localized lesions can have good outcomes with repeated treatment. However, extensive AVMs, involving the entire extremity, are difficult to treat, and they usually have unfavorable outcomes.

To reduce the ethanol-related local and systemic complications, several sessions of embolotherapy were required in most patients. The interval between sessions is generally 3 months to 6 months, depending on patient symptoms or the extent of AVMs. Park et al. (9) reported that with experience, the number of treatment sessions can be reduced to almost half (4.2 sessions in the first decade and 2.3 sessions in the last decade), and treatment failure can be reduced significantly from 9.4% within the first decade to 1.5% within the last decade. This outcome may be attributed to the growing knowledge of AVM hemodynamics and the modifications of the treatment technique based on the angiographic types of AVMs (9).

The reported complication rates of ethanol embolotherapy were 23% overall, 20% for minor complications, and 3% for major complications. Skin necrosis, bullae formation, and transient nerve injuries were the most common minor complications (4). For the major complications, skin necrosis that required skin grafts, amputation for tissue necrosis, thrombolysis for distal embolism, permanent nerve injury, acute pancreatitis from ethanol use, and acute renal failure were reported. No procedure-related mortality was reported.

**Special Consideration**

**Hand and Foot AVMs**

Patients with hand and foot AVMs have more severe ischemic pain, ulceration, and/or necrosis. The treatment outcomes for hand and foot AVMs are relatively poor; they have lower cure and higher complication rates than AVMs in other locations. Hand AVMs involving the fingers had relatively more complications than those without
finger involvement. If an AVM is small, it is difficult to puncture or catheterize the nidus or discriminate between AVM and normal vessels on angiography. Moreover, the risk of inevitable embolization or reflux to normal vessels increases, and coil embolization to reduce the amount of ethanol is difficult due to the narrow space and the superficial location. To reduce the complications, meticulous discrimination and infusion techniques that reduce the amount and concentration of ethanol are required (2, 17).

Bone AVMs

Intraosseous AVMs of the extremities have been associated with distortion, hypertrophy, osteolytic skeletal changes, leg-length discrepancies, and pathologic fractures. When treating bone AVMs, cortical erosion or thinning, which is an accessible route for direct puncturing, is key to the treatment approach. When cortical erosion or thinning is adjacent to the nidus of the bone AVM, a direct puncture can be performed. When there is no accessible route for direct puncturing due to a thick cortex, the angiographic classification of the AVM can be considered. A transvenous approach for a type II bone AVM and a transarterial approach for a type III bone AVM are recommended for embolotherapy.

A pure bone AVM is confined to the bone, whereas mixed bone and soft-tissue (MBS) AVMs involve the skin, subcutaneous fat, or muscle in addition to the bone. The clinical success rate of pure bone AVMs is higher than that of MBS AVMs due to the skin and subcutaneous tissue component of MBS AVMs. Because the treatment of these skin and subcutaneous AVMs can result in severe skin necrosis, compression stocking, instead of further embolotherapy, is recommended to relieve swelling and pain for the residual skin and subcutaneous components of MBS AVMs (18, 19).

CONCLUSION

By using different treatment strategies for individual types of AVMs, ethanol embolotherapy with or without coils induced a high cure rate with acceptable major and minor complications. The angiographic classification and treatment strategy for peripheral AVMs may guide therapeutic teams in planning the optimal therapeutic approach based on a better understanding of the characteristics of AVMs in the body and extremities.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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