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REVIEW ARTICLE

# Interdisciplinary management of peripheral arteriovenous malformations: review of the literature and current proceedings

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**ABSTRACT**

Arteriovenous malformations (AVMs) are a rare congenital vascular disorder. They represent a fast-flow vascular malformation. Clinically, AVMs present a heterogenous expression and can affect every part of the body. Here, we will solely focus on extracranial AVMs. Generally, AVMs progress with the patient's age. Patients often suffer from pulsation, skin discoloration, pain, ulceration, bleeding, and disfigurement. Diagnostic tools include color-coded duplex sonography, MRI and CT imaging, as well as the clinical examination. 4D dynamic perfusion-computed tomography may help in the interventional planning. Digital subtraction angiography is required during interventional therapy. AVMs pose a great challenge to the treating physician. The therapy of this rare disease should be managed in an interdisciplinary center for vascular malformations. It consists of conservative measures, such as compression garments and pain medication, transcatheter or, more rarely, percutanous embolization, and surgical resection. In smaller, localized lesions, resection with primary wound closure may be feasible, whereas extensive AVMs regularly require the reconstruction of the resulting soft tissue defect and possibly affected functional structures by means of free tissue transfer. In the interdisciplinary setting required for an appropriate treatment of AVMs, extensive knowledge of the various therapies, including those from different specialties, is necessary. Therefore, this article aims to provide an overview over both the interventional and surgical therapeutic options.

**Abbreviations:** AVM: arteriovenous malformation; CT: computed tomography; DSA: digital subtraction angiography; GLUT1: glucose transporter 1; 4D: 4-dimensional; MAP2K1: mitogen-activated protein kinase kinase 1; MRI: magnetic resonance imaging; MR-A: magnetic resonance angiography; mTOR: mammalian target of rapamycin; RASA1: RAS p21 protein activator 1; RNA: ribonucleic acid; SMAD4: mothers against decapentaplegic homolog 4

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Arteriovenous malformations; vascular anomaly; vascular malformation; interventional radiology; embolization; reconstructive surgery

## Introduction

Vascular anomalies are classified according to the 2018 ISSVA classification [1]. Arteriovenous malformations (AVMs) represent fast-flow vascular anomalies with heterogenous clinical manifestation. AVMs occur with a prevalence of 1.5–4.5% and an incidence of 1 per 100,000 capita per year and are classified as rare diseases [2]. The cause of AVM development is thought to be an error in vasculogenesis.

Several genetic mutations have been identified to cause hereditary AVMs. A deficiency of *SMAD4* has been shown to result in formation of AVMs in mice [3]. Furthermore, an association of mutations in the *MAP2K1* gene has been demonstrated [4]. Mutations in the *RASA1* gene may be associated with the Parkes–Weber Syndrome [5].

AVMs pose a great challenge to the treating physician. They generally progress with the patient's age and may cause pain, disfigurement, ulceration, bleeding and even high-output cardiac failure [6]. Treatment should be managed by an experienced multidisciplinary team of physicians, including interventional radiologists, pediatric surgeons, vascular surgeons, hemostaseologists, craniomaxillofacial surgeons, otolaryngologists, general and thoracic surgeons and also reconstructive microsurgeons. Concise treatment planning is very important, since insufficient or inappropriate treatment may significantly worsen symptoms and outcomes. Clinically, AVMs have long been staged according to the Schobinger classification [7] (see Table 1).

One major problem in the treatment of AVMs lies in their scarcity. Among many physicians, vascular malformations are yet a not very well-known entity and, therefore, may be falsely diagnosed or treated [8]. Early diagnosis and treatment of a symptomatic vascular anomaly is crucial to provide good clinical outcomes and patient satisfaction.

In this article, diagnosis and therapy of fast-flow vascular anomalies are presented and discussed with regard to the current literature. The clinical characterization of the lesions, diagnostic imaging spectrum and treatment possibilities are described to facilitate a sound assessment of fast-flow AVMs. We aim to

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demonstrate these complex findings by presenting patients that have undergone interdisciplinary management in our institutions. However, this review of the literature does not intend to be a case series and, thus, refrains from presenting the entirety of each case.

## Materials and methods

We reviewed the literature on Pubmed for all citations that identified vascular anomalies with regard to arteriovenous malformations from 1970 until 2019. Search terms included 'vascular anomaly', 'arteriovenous malformation', 'arterio-venous malformation', 'therapy', 'diagnosis', 'diagnostics', 'hemangioma', 'interventional therapy', and 'surgical therapy'.

The cases that are reported in this manuscript are based on regular clinical data that have been derived from clinical consultation and treatment of the patients in a vascular anomaly center. No further interview or contact was conducted. No third parties had access to any patient-related data during the conduction of this study and all data that were stored were previously anonymized. Therefore, no institutional review board was required according to the local legislation.

## Diagnostics

Thorough clinical examination and acquisition of a detailed medical history are key factors in the diagnosis of a vascular anomaly. Imaging completes the information on flow characteristics of vascular anomalies, like for example AVMs.

Clinically, AVM patients may experience local symptoms like a warm and reddish skin in proximity of the lesion, pulsation, pain, and ulcerations with bleeding, but also cardiac failure (see Table 1). AVMs may be quiescent until the onset of puberty, which may be related to the expression of estrogen and progesterone receptors in AVMs [9,10]. Trauma may induce rapid progression [11]. In cases of localized intravascular coagulopathy, patients exhibit elevated D-dimer levels and often times experience amplified pain [12].

Table 1. Schobinger classification of arteriovenous malformations.

| Stage | Clinical findings |
|---|---|
| I – Dormancy | Local skin hyperthermia, red skin coloration |
| II – Expansion | Enlargement, pulsation, bruit |
| III – Destruction | Skin alteration, bleeding, necrosis, pain |
| IV – Decompensation | Stage III + high output cardiac failure |

By means of color-coded duplex ultrasonography, fast-flow patterns with hypervascularity and arterio-venous shunting can be identified (see Figure 1) which are characteristic for these vascular anomalies. One has to keep in mind, that in gray-scale imaging, the lesion may not be visible to its full extent [13].

In suspected AVMs, the diagnosis may be confirmed on ultrasound. For precise assessment of size and extension of the vascular anomaly, MRI is recommended. MR-angiography enables vascular characterization of the fast-flow dynamics of AVMs, especially with focus on feeding arteries, the nidus, and draining outflow veins. On post-contrast T1-weighted sequences, the involvement of soft tissue, muscle, and bone can be assessed further. AVMs usually do not demonstrate an extensive soft tissue enhancement. One of their typical characteristics on T2-weighted spin echo sequences in MRI is the presence of flow voids, caused by signal loss due to the presence of bundles of blood-filled vessels surrounding the nidus of the AVM. This imaging finding differentiates AVMs from vascular tumors which usually appear as solid and compact hypervascularized lesions in post-contrast T1-weighted imaging (see Figure 2(A,B)) [14].

Inflow and outflow vessels of an AVM may be identified using TWIST MR-Angiography (time-resolved angiography with interleaved stochastic trajectories). This is a time-resolved technique with very high temporal resolution, which is essential for the analysis of fast-flow vascular anomalies [15]. TWIST MR-A allows rapid acquisition of multiple images over time during the passage of the injected contrast bolus, enabling the evaluation of the blood flow through the AVM.

The MRI protocols for fast-flow vascular anomalies should include multiplanar T1 TSE sequences, multiplanar T2 TSE

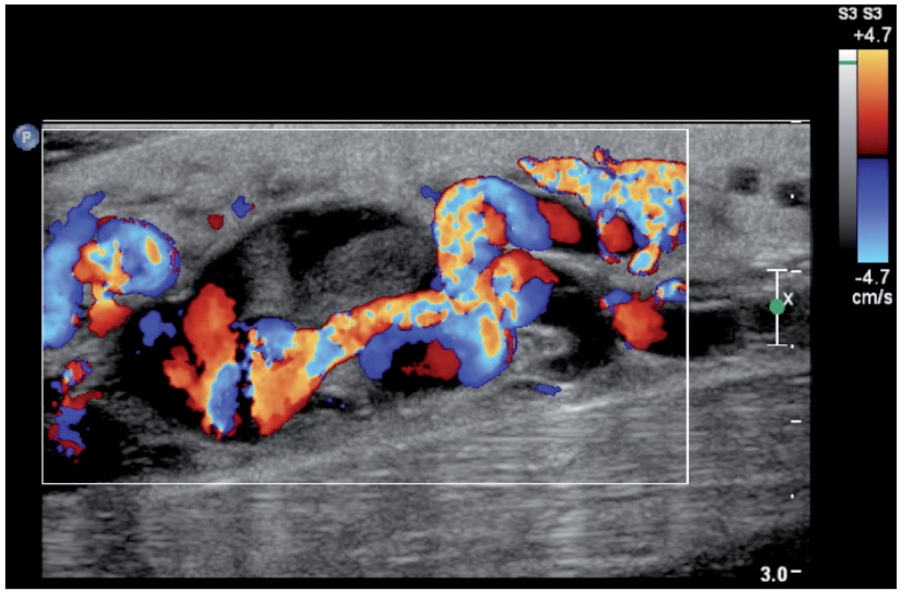


Figure 1. Color-coded duplex ultrasonography demonstrates a fast-flow pattern with hypervascularity and arteriovenous shunting in a peripheral AVM of the lower limb in a patient with dilated and tortuous vessels; this 17-year-old patient suffered from an AVM of the thigh. Complete nidus occlusion was achieved in two embolization sessions (timed 6 months apart). Postinterventionally, the patient had persistent pain and disliked the bulk in his thigh and thus, surgical resection was performed six months after the embolization (same patient as in Figures 4 and 5, image taken before the embolization).

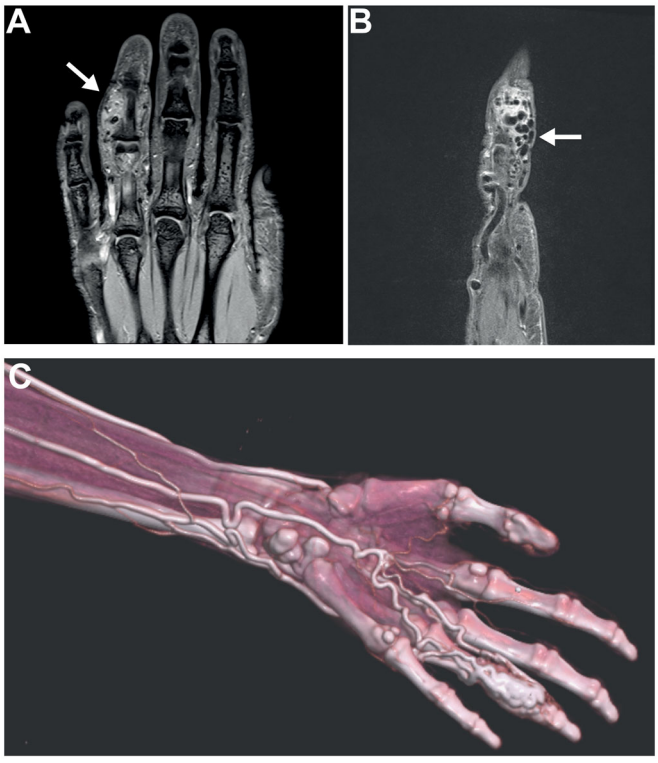


Figure 2. MRI and 4D CT imaging of AVM of the right ring finger; (A) post-contrast T1-weighted MRI in coronal view with and digital AVM involving digit IV and contrast uptake in the soft tissue mass surrounding the vascular anomaly, (B) the sagittal view of digit IV, and (C) 4D perfusion CT of the AVM of digit IV on a SOMATOM Force CT scanner (Siemens Healthineers, Erlangen, Germany). The anterior interosseous artery supplies the AVM with demarcation of the dilated nidus at the middle phalanx of digit IV.

sequences, time-resolved 3D MR-angiography and early post contrast T1-weighted fat saturated sequences. Despite potentially prolonged examination time, MRI should always be the first imaging choice to obtain an overview on AVM flow dynamics. It does not expose the patient to radiation, which is a major point to be considered in the usually young patients.

In very extensive or multisegmental AVMs, delineation of AV shunts to and from the nidus can be disguised in case of inappropriate bolus timing. We recommend 4D dynamic perfusion computed tomography with high spatial resolution for the preinterventional assessment of extensive AVMs (see Figure 2(C)) [16]. Thorough diagnostics of the manifold arterial and venous shunts and configuration of the AVM nidus before embolization has tremendous impact on intervention planning. Vascular access, selection of catheters, and choice of embolic agents are facilitated along with intraprocedural reduction of X-ray exposure [16].

Digital subtraction angiography (DSA) is usually performed during interventional treatment and is not recommended as a stand-alone diagnostic procedure. For the interventional treatment, transarterial or transvenous angiography and occasionally also percutaneous access after direct puncture of the AVM may be needed to enable efficient embolization (see Figure 3(A)). Cho et al. proposed an angiographic classification based on the vascular anatomy in DSA findings in 2006 [17], which has come to be widely accepted as the standard angiographic classification (see Table 2).

Colleti et al. proposed a new staging system for AVMs of the head and neck in 2020, which was established in a multinational, multicenter setting [18]. Surgical/anatomical, endovascular, and clinical features as well as growth of the AVM are integrated into a staging system. Colleti et al. reported to already have successfully implemented the classification in a series of more than 100 patients. By including both surgical and endovascular aspects into a single classification, the new SECg staging system might be well suited for guiding the therapy of AVMs. With minor adjustments, it may be used in the staging of trunk or extremity AVMs, too. However, further evaluation of this new tool is necessary. The SECg staging system is depicted in Table 3.

Histologically, AVMs represent abnormal direct connections between arteries and veins skipping the organ-specific capillary bed. These abnormal arterio-venous communications consist of arterioles, capillaries, and venules haphazardly aggregated into vascular clusters. Abrupt dilation and changes in the vessel wall thickness or structure are typical findings. The veins often show reactive intimal hyperplasia. The adventitia and the adjacent connective tissue are fibrosed (Figures 4(D–F) and 7(G)). Depending on the localization and patient history, the lesions may show hemorrhage, necrosis and ulceration of the skin or mucosa. The pathologist is commonly confronted with the differential diagnosis of hemangiomas. In contrast to AVMs, hemangiomas typically lack arterial and arteriolar structures and intralesional nerve fibers [19]. Clinically, hemangiomas would present as solid, bright red, hypervascularized tumors, densely packed with blood vessels. Histopathologically, they show positivity for GLUT-1 in their endothelium [20].

## Multimodal therapy

The treatment of fast-flow vascular anomalies is challenging and consists of a multitude of therapeutic approaches which should always be discussed and conducted by an interdisciplinary team of experts in their respective medical fields.

### *Conservative treatment*

AVMs in Schobinger Stage I may be treated solely on a watch-and-wait basis. Compression garments can help to reduce symptoms like swelling, pain, and hyperhidrosis. Analgetic and antiphlogistic medication should be considered if needed. Female patients seeking birth control should consider oral contraceptives with reduced estrogen content or alternative contraception methods [15], since elevated estrogen levels during pregnancy or under oral contraception may induce stimulation of the AVM and potential disease progression [10,11,19,21–23]. However, it has to be noted that there have not been prospective trials comparing oral contraception with other methods of contraception in this patient population and this recommendation, therefore, is mainly based on eminence, not evidence.

In cases of associated localized intravascular coagulopathy, patients might require anticoagulation and should be monitored by a hemostaseologist as well. Regular consultations of these patients are recommended to detect progression or worsening of symptoms in an enlarging AVM that will eventually require therapeutic intervention. As AVMs increase with the patient's age, worsening of the current findings is to be expected and patients should be considered for interventional therapy in Schobinger stage II.

### *Pharmacotherapy*

Recently, mTOR inhibitors, such as Sirolimus/Rapamycin, have been tested in clinical trials in an effort to prevent progression and reduce the rate of recurrence in various vascular malformations [24–26]. Gabeff et al. observed a partial response in 5 of 10

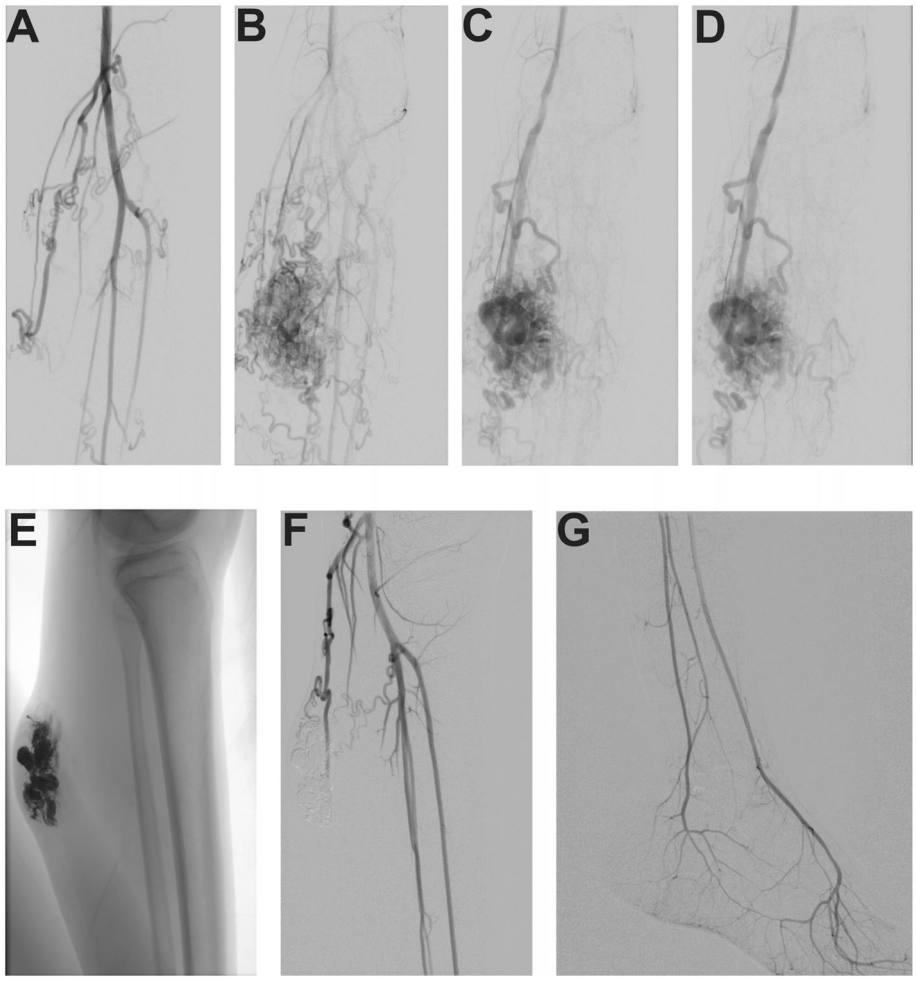


Figure 3. Pre-interventional catheter angiography of the left lower limb of the patient in Figures 1 and 5, catheter angiography prior (A–D) and after embolization with Onyx (E–G). The nidus of the AVM is completely occluded after embolization with rapid contrast material run-off into the peroneal arteries and patent arterial supply to the foot.

Table 2. Cho-classification of DSA findings in AVM [17].

| Anatomical type | Angiographic findings |
|---|---|
| Type I | Arteriovenous fistula:<br>No more than 3 arteries shunt into a singular draining vein |
| Type II | Arteriolovenulous fistula:<br>Multiple, plexiform arterioles shunt into a singular draining vein |
| Type IIIa | Arteriolovenulous fistulae with non-dilated fistulae:<br>Multiple fine shunts between arterioles and venules, appearing as a blush |
| Type IIIb | Arteriolovenulous fistulae with dilated-fistulae:<br>Multiple shunts between arterioles and venules, appearing as a vascular network |

AVM patients treated with Rapamycin in a retrospective analysis, but also reported therapeutic resistance after three months in two of those five patients [27]. Sandbank et al. [28] found improvement in discoloration, appearance and pain in one patient. Six patients suffering from AVMs were treated with oral Sirolimus and embolization or surgical debulking by Chelliah et al. in 2018 and responded favorably according to the authors [26]. In a retrospective study by Triana et al., four AVM patients showed no response to oral Rapamycin [25]. Furthermore, various other anti-angiogenic drugs as well as propranolol have been tested and even used successfully in single patients [29–31]. Nonetheless, more scientific evidence is needed in the field of pharmacotherapy for vascular malformations.

### *Minimally invasive interventional therapy*

Diagnostic angiography and/or venography is the prerequisite for the therapeutic embolization of AVMs, either as a presurgical or definitive procedure. Embolization of AVMs often is challenging and requires interventional experience by the radiologist who should be very familiar with embolic agents and catheter devices. These procedures should be carried out in dedicated vascular anomaly centers.

Transcatheter embolization may be indicated as definitive therapy of peripheral AVMs. It is also applied as a preoperative tool for debulking and size reduction of the vascularization of the AVM with the intention of decreasing the risk of excessive blood loss during surgical resection. Preoperative embolization should be performed within a few days prior to surgery to avoid formation of potential collaterals that might stimulate growth and recurrence of the AVM [15,32].

The goal of an embolization must be the total occlusion of the nidus [33–35]. The nidus consists of densely packed, tortuous blood vessels with a very low vascular-resistance tonicity, thereby causing recruitment of collateral inflow and arterialization of the venous system [33,36,37]. Most commonly, liquid casting agents, such as ONYX® or SQUID® (ethylene vinyl alcohol copolymers),

**Table 3.** SECg staging system for AVMs of the head and neck by Colleti et al. [18].

| Stage | Surgical/anatomical | Endovascular | Clinical | Growth |
|---|---|---|---|---|
| 0 | – | – | No symptoms | – |
| 1 | Involvement of a single anatomical site | ArterioVenous AVM | Symptomatic, but without complications | – |
| 2 | Involvement of two adjacent anatomical sites | ArterioloVenous AVM | Local complications | – |
| 3 | Infiltration of important, but not vital structures | ArterioloVenular AVM | General Complications | – |
| 4 | Infiltration of vital structures | – | – | – |
| – | – | – | – | Stable during last 6 months |
| + | – | – | – | Progressive during last 6 months |

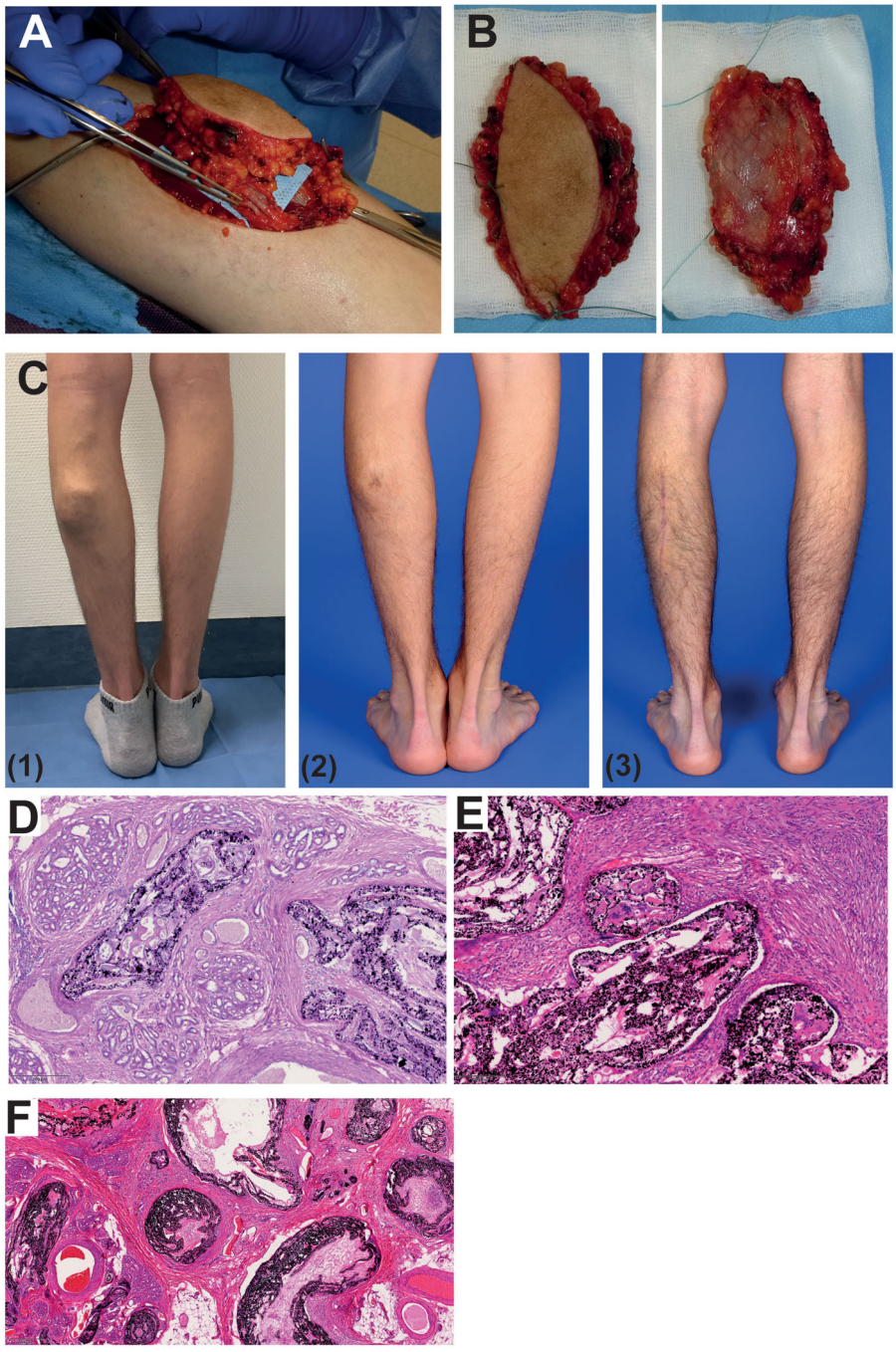


**Figure 4.** Clinical and histopathological aspects of the same patient as in Figures 1 and 4; (A) intraoperative view of the large draining veins exiting the AVM, after resection primary closure with intracutaneous suture was feasible, (B) surgical specimen after complete resection, (C) clinical aspect of the AVM of the left thigh, before (1) and after embolization (2), after surgical nidus resection (3); (D, E) histopathology: higher magnification showing the full-blown fibrosing processes initiated by the embolization. Foreign-body type giant cells can be readily recognized in panel (D); (F) overview showing atypical vessels of different sizes localized in clusters. The embolized material stains black; staining: E + F: HE, D: PAS.

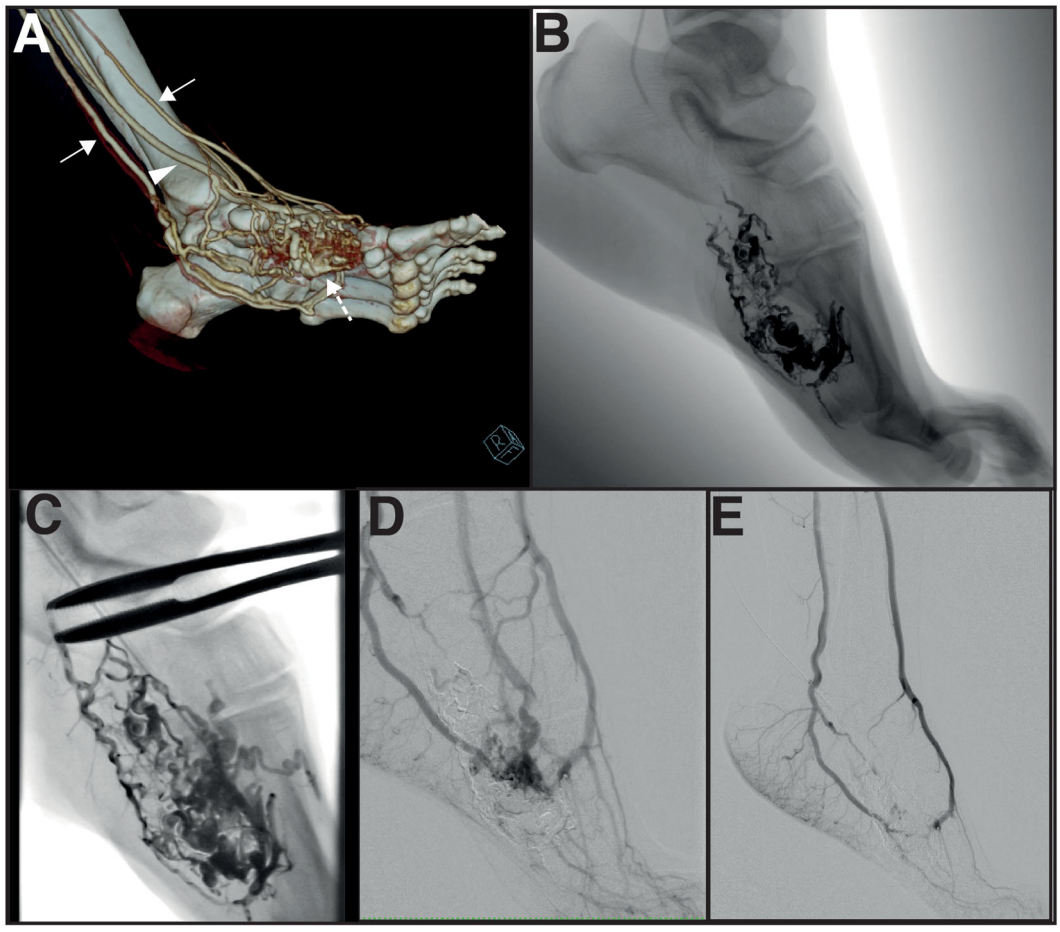


Figure 5. Dynamic 4D perfusion CT with postprocessed sagittal view of a plantar AVM of the left foot (solid arrows). Arterial supply from the tibial anterior and posterior artery (dashed arrow) and a dominant outflow vein (arrowhead) (A). Anatomical overview after transarterial embolization of the pedal AVM with ethylene vinyl alcohol copolymer (B) and digital subtraction angiogram still demonstrating patency of the AVM, mainly through venous inflow (C). Percutaneous approach to the AVM with a puncturing needle (D) and continuation of the embolization until final overview angiography demonstrates total nidus occlusion (E).

are used in a push and plug technique (see Figure 3) [35]. Ethylene vinyl alcohol copolymers feature a good safety profile. They have been in use for intracranial embolization for several years and exhibit good permanent occlusion properties [38–40]. Yet, their high costs have to be considered and weighed against its effective embolizing characteristics and safety profile compared to particulate and other liquid embolics. In fast-flow lesions, ethanol or other direct sclerosants may also be applied, but increased care should be taken to avoid washout of the sclerosant due to the very high flow rates in AVMs and potential irreversible nerve damage after ethanol embolization [34].

In all embolization therapies, patients have to be monitored for local complications like migration of the embolization agent during the procedure, resulting in non-target infarction and ulceration or potential nerve injury [35]. In cases of incomplete nidus occlusion, AVM recurrence rates are extremely elevated and have been reported to be as high as 98% [19].

In patients with extensive AVMs, a transarterial route to the nidus may not be expedient due to the tortuosity of inflowing arteries, which are fragile and can dissect easily. In these cases, a combined approach with transarterial, transvenous, and direct percutaneous embolization should be considered. Especially, the dominant outflow vein can be targeted with a percutaneous retrograde approach to completely occlude the AVM nidus (see Figure 5).

In large AVMs with strongly dilated outflow veins, the transvenous retrograde push-through embolization technique is recommended for total nidus occlusion (see Figure 6). In this procedure, a protection device like an occlusion balloon catheter or Amplatzer plug is temporarily placed in the proximal part of the dominant outflow vein to prevent migration of the liquid embolic agent into major central veins causing pulmonary artery embolism.

Then, the embolic agent is pushed retrogradely through the veins into the arterial part of the AVM.

### *Surgical therapy*

Surgical therapy plays a prominent role in the management of fast-flow vascular anomalies, but is less common in the management of slow-flow lesions like venous malformations.

Among many aspects, size, localization, and symptoms of the AVM, as well as the patient's age need to be considered. Thus, the decision for surgical intervention has to be made individually for each patient. It is generally accepted that the resection of a fast-flow vascular malformation must not leave the patient with a deformity, which is worse than the original anomaly. This is especially important in cases of involvement of the facial region or the hands, where extensive resection may be feasible, but might lead to an aesthetically or functionally unfavorable outcome.

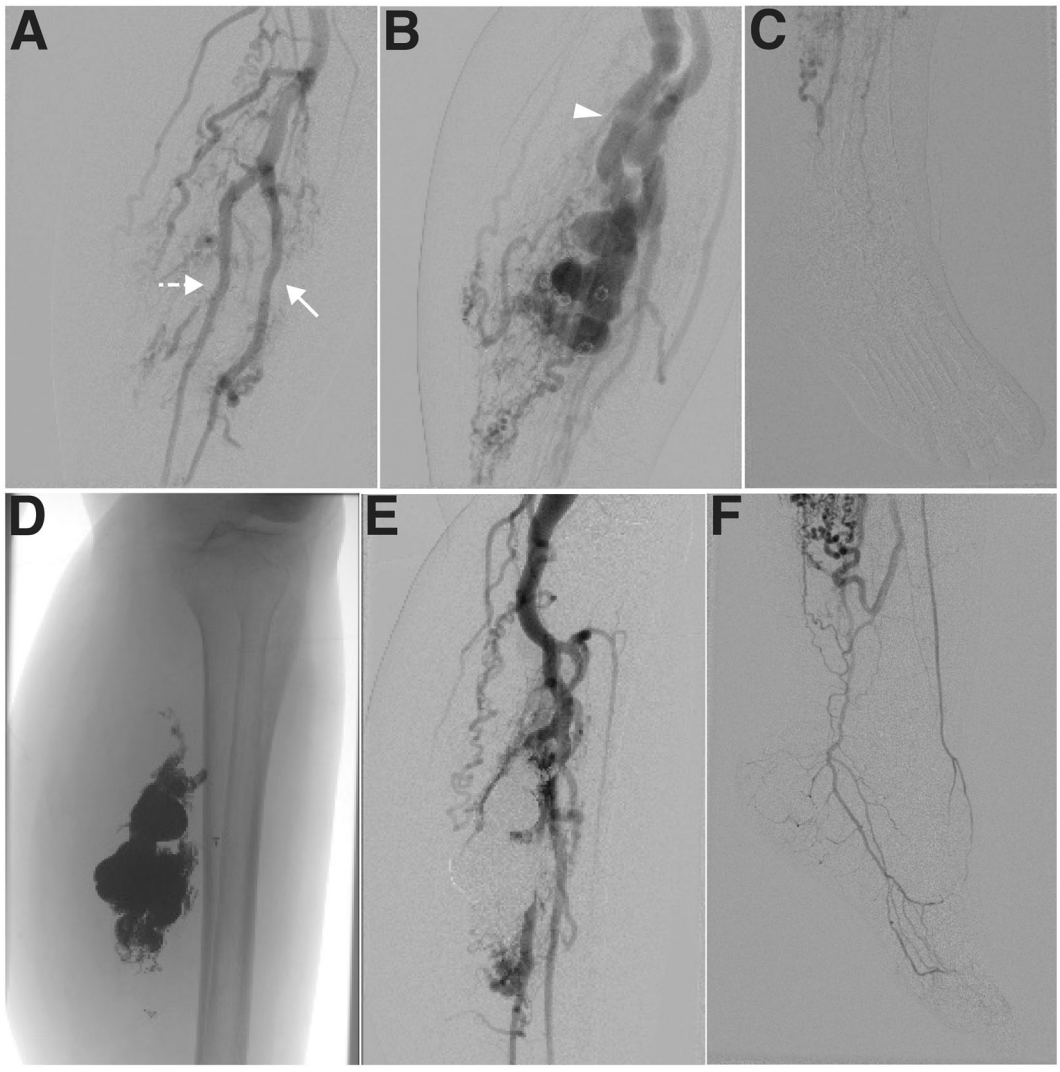


**Figure 6.** Transarterial catheter angiography in early phase demonstrates the anterior tibial (arrow) and peroneal artery (dashed arrow) (A) with dilated dominant outflow veins (arrow head) draining the nidus of the AVM (B). Angiogram does not demonstrate patency of the dorsalis pedis artery (C). Percutaneous transvenous push through embolization of the AVM nidus in the calf with ethylene vinyl alcohol copolymers, pushing the embolic agent retrograde into the arterial feeder of the AVM (D). Catheter angiography demonstrates a not yet complete devascularization of the AVM (E). At this point of the embolization, reduction of AVM shunts has increased contrast inflow into the dorsalis pedis artery (F).

**Table 4.** Revised Richter-Suen classification system for extracranial arteriovenous malformations [31].

| Focal AVM | Multicentric (diffuse) AVM |
|---|---|
| Central nidus | Nidus difficult to detect or multiple |
| Single arterial feeder | Two or more arterial feeders |
| Firm | Compressible, rapid rebound |
| Singular skin involvement, if present | Diffuse skin involvement in affected region |

Recurrence rates after resection are reported to be lower than after interventional therapy alone [19,41]. Due to the progressive nature of AVMs, early therapy decision should be considered, since surgery in later stages of the disease might be much more extensive and challenging [26,32,42,43]. Richter and Suen published a classification of arteriovenous malformations, which was revised by Uller et al. in 2014 (see Table 4) [32]. It differentiates between focal and multicentric AVMs.

In smaller, focal AVMs, an excision with primary closure is often feasible and should be considered primarily (see Figure 4(A–C)). Depending on the AVM size, preoperative embolization may be advisable to minimize blood loss. Furthermore, the liquid embolic cast with its demarcation of the embolized area, which turns grey or black in color due to the radiopaque tantalum [44], may help the surgeon in guiding the dissection. It should be noted that the cast is highly combustible and may cause sparks when in contact with monopolar cautery. Excision should optimally be planned 24–48 h after the embolization, before revascularization of the AVM through neighboring vessels and local inflammation due to the foreign body reaction to the cast occur [15,32,41,43].

In cases of extensive resection and inclusion of functional structures, such as tendons, blood vessels or nerves, reconstruction of the resected structures has to be performed (see Figure 7(A–F)).

More advanced focal lesions and diffuse lesions usually cannot be resected without reconstruction of the resulting soft tissue defect. Local flaps and microsurgical tissue transfer should be considered in these patients (see Figure 8). In few cases of very large fast-flow vascular malformations, especially in the head and neck

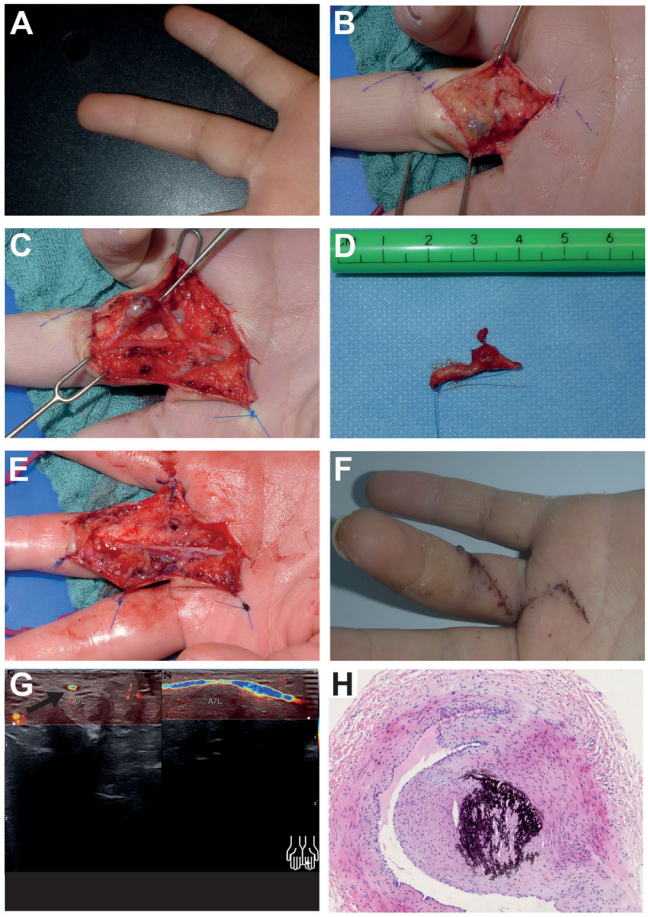


Figure 7. A 15-year-old patient with an AVM of the seventh digital artery (A7) on the left ring finger. Due to the small size of the lesion and the possibility of using a tourniquet, the decision not to perform preoperative embolization was made in an interdisciplinary board meeting; (A) pre-operative clinical aspect, (B) and (C) intraoperative view of the AVM after Bruner incision, (D) surgical specimen after complete resection, (E) intraoperative aspect after reconstruction of A7 *via* subcutaneous vein graft from the palmar forearm, (F) clinical aspect on POD 14 after removal of stitches; (G) color-coded duplexsonography showing patency of the interpositional vein graft for digital artery 7 and 9 months postoperatively; (I) histopathological overview showing atypical vessel with an irregular endothelial architecture and prominent intimal hyperplasia (HE staining).

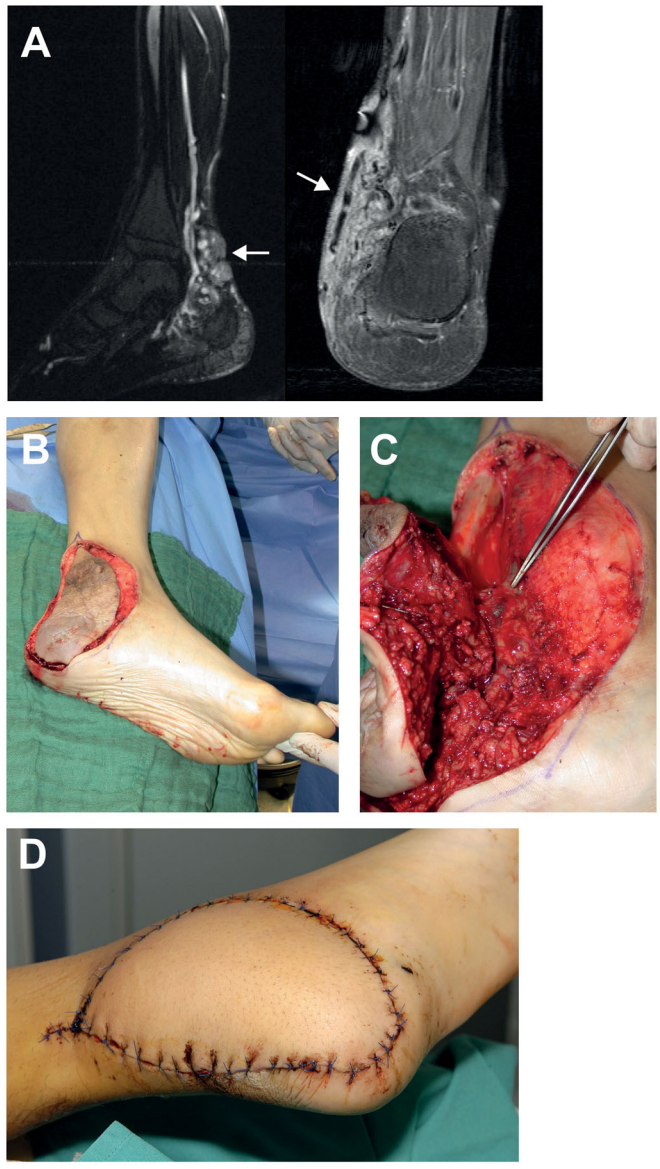


Figure 8. A 26-year old patient suffering from an AVM of the medial ankle. Prior to presenting in our institution the patient had been embolized three times (6, 5, and 3 years before surgery), but no significant reduction of size and symptoms had been achieved. Further embolization had been deemed unfeasible and thus no preoperative embolotherapy was undertaken; (A) preoperative MRI, (B) intraoperative aspect of the AVM, skin alterations due clearly visible, (C) feeding vessels exposed before complete excision, (D) postoperative aspect of antero lateral thigh (ALT) flap.

region, complete resection might not be feasible at all, if the functional or aesthetic defect after complete resection would be inacceptable. Staged resection with the goal of completely resecting the nidus or a palliative approach to reduce symptoms are indicated then [15,32,41]. It has to be noted that with significant progresses in reconstructive (micro)surgery, complete resection of the majority of high-flow malformations is feasible and incomplete resections due to limited availability of advanced reconstructive microsurgical armamentarium are not acceptable.

Greene et al. reported that split or full thickness skin grafts have a high failure rate, if transplanted into ulcerated areas [15]. It is hypothesized that due to the high shunting volume in fast-flow vascular malformations, the surrounding tissue suffers from ischemia. Therefore, skin grafting should only be used with caution and in selected cases.

The suggestion that wide resection margins reduce recurrence rate has not been proven sufficiently. Thus, we suggest a complete resection with the goal of entirely removing the nidus, while preserving as much unaffected tissue as possible. Skin coloration and bleeding from the wound edges may help in determining the necessary resection margins [11].

Ligation of feeding arteries without attempting a complete resection has been proposed in the past, but nowadays proximal ligation has been shown to rather increase the risk of progression and should not be attempted [11,15,45]. Incomplete resection of the nidus will always lead to a recurrence of the AVM with progression of the disease.

The following general principles may aid the surgeon in reducing intraoperative blood loss and enabling a straight forward procedure (adapted from Uller et al. [32]):

1. Use of a tourniquet in extremity lesions.
2. Infiltration of epinephrine containing local anesthetic (i.e. 1:100,000 dilution) in the surgical field if a tourniquet is not applicable.
3. Allowance of appropriate operating time, since the resection might need prolonged hemostasis.

4. Availability of two sets of monopolar and bipolar cautery, attached to two separate cautery machines for extensive AVMs.
5. Preoperative preparation of erythrocyte concentrates for patients with extensive lesions.
6. Placement of suction drains.
7. Contact a hemostaseologist prior to surgery in case of suspected coagulation disorder.

Postoperative wound dehiscence and wound healing disorders are frequently encountered [32,43]. Close patient monitoring by frequent outpatient follow-up is advised.

Recurrence rates are high, even after a macroscopically complete resection. Rates of up to 85% have been reported [19]. Koshima et al. promote radical resection and reconstruction by free tissue transfer [46]. More recently, Visser et al. reported a recurrence rate of only 8.7%, when differentiating between a curative and a palliative approach [41]. Even if complete resection is not possible, patients may benefit from symptom reduction after partial resection [11,15,19,41].

In many patients, embolization therapy may be sufficient to control AVM symptoms. In cases where interventional radiological procedures alone are not sufficient, surgery decreases the recurrence rate and supports an improved functional and cosmetic outcome.

## Conclusion

Diagnosis and treatment of AVMs require extensive knowledge of this rare disease. An interdisciplinary approach involving different clinical specialists is required to establish a dedicated treatment plan that will enable optimal postprocedural results. To ensure the best possible treatment, we strongly urge every treating physician to refer patients with fast-flow vascular malformations to dedicated centers for vascular anomalies.

## Disclosure statement

No potential conflict of interest was reported by the author(s).

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