Vascular Malformations: An Update on Classification, Clinical Features, and Management Principles

Sumit R Kapadia, Vijay M Thakore, Hiten M Patel
Department of Vascular and Endovascular Surgery, Venus Hospital, Department of Vascular and Endovascular Surgery, VINS Hospital, Vadodara, Gujarat, India

Abstract

Congenital vascular malformations are one of the most challenging subgroup of diseases treated by vascular surgeons and interventionalists. Currently, there exists a lot of misunderstandings and controversies in terminology, diagnosis, and management of patients with these problems. This review article helps doctors with a concise and current understanding of classification, clinical features, complications as well as diagnostic and therapeutic guidelines.

Keywords: Arteriovenous malformation, embolization, Klippel–Trenaunay syndrome, sclerotherapy, vascular malformations

Introduction

The aim of this article is to present a review of the different types of vascular anomalies including malformations including classification, identification, clinical features, diagnostic criteria as well as management principles.

Vascular anomalies were classified originally by Mulliken and Glowacki in 1982 based on clinical features and biologic behavior. However, currently, the International Society for the Study of Vascular Anomalies (ISSVA) classification system is widely accepted and utilized to categorize vascular anomalies into two basic types: (1) vasoproliferative or vascular neoplasms such as hemangioma and (2) developmental vascular abnormalities called congenital vascular malformations (CVMs).

The clinical importance of classification is important to reduce the confusion in treatment principles.

Hemangiomas are one of the most frequent tumors of infancy and can be classified as the common infantile hemangioma (typical angioma) or the rare congenital hemangioma (rapidly involuting congenital hemangioma or noninvoluting congenital hemangioma). Infantile hemangiomas classically present between 2 weeks and 2 months of age.

Clinically, a superficial hemangioma appears as a skin mark or mass with a typical cherry-red color [Figure 1]. The diagnosis of such hemangiomas is straightforward and may not warrant any investigation. Most hemangiomas also do not require immediate intervention and undergo spontaneous involution. While rapid involution is noted in 50%–60% of children by the 2nd year of age, almost 90% can be expected to undergo gradual involution before the age of 9 years. In 50% of these patients, normal skin is restored, whereas in the remaining, the cosmetic result is much better than that offered by surgical intervention. Review articles also suggest that involution of 50%, 70%, and 90% of the hemangioma occurs by five, seven, and 9 years of age, respectively, with some discrepancy between reports. At the final stages of involution, a fibrofatty protuberance may remain.

Systemic or intralesional steroids as well as systemic propranolol (in dose of 1–3 mg/kg/day) and topical timolol have been found to be useful in reducing size of infantile hemangiomas, especially near vital areas or with complications. [Figure 2a and b] There have been reports of usefulness of interferon alpha 2a for large hemangiomas as it blocks migration and proliferation of endothelial cells, smooth muscle cells, and fibroblasts by decreasing the production of collagen and basic fibroblast growth factor.

Address for correspondence: Dr. Sumit R Kapadia, E-mail: sumit.kapadia@gmail.com

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Kapadia SR, Thakore VM, Patel HM. Vascular malformations: An update on classification, clinical features, and management principles. Indian J Vasc Endovasc Surg 2017;4:152-62.

Received: September, 2017. Accepted: September, 2017.
The dosage used is $3 \times 10^6$ U/m$^2$/day subcutaneously for 6–18 months.\(^7\)

Hence, almost no vascular birthmark present in an adolescent or adult should be described as a hemangioma; it may actually be a vascular malformation. The differences between hemangioma and vascular malformations are enlisted in Table 1.

CVMs are a large group which include venous malformation (VM), arterial malformation, capillary malformation (CM), and lymphatic malformation (LM), as well as arteriovenous malformation (AVM) malformations. They remain a difficult diagnostic and therapeutic challenge due to the varied clinical presentations, unpredictable clinical course, inconsistent response to treatment with high recurrence rates, and confusing terminology.\(^8,9\)

**Classification of Congenital Vascular Malformation**

Earlier, different classifications have been used, based on anatomy, histology, physical appearance, or development of the malformation. In addition, flow velocity – high or low – in the malformation was used to distinguish one lesion from another. The Hamburg classification uses vessel type as the basis of classification of vascular malformations. In each class, “truncular malformations” affecting (individual) large vessels and “extratruncular malformations” composed of smaller vessels intimately embedded in the host tissue are recognized.\(^10\)

An updated version of the ISSV A classification has been laid down in 2014 in which CVM are divided into 4 groups: simple malformations, combined malformations, malformations of major named vessels (similar to truncal malformations), and malformations associated with other anomalies (most of which are the eponymous syndromes).\(^11\) Many of these malformations become apparent during puberty because they have hormone receptors that make them sensitive to estrogen and testosterone variations.

**Clinical Features of Congenital Vascular Malformation**

**Venous malformations**

VMs occur in vessels that are morphologically and histologically similar to veins and hence have low blood flow and are compressible. They are categorized as either superficial or deep, and as localized, multicentric, or diffuse.\(^12\) The appearance of most superficial ones is purple color [Figure 3]; the subcutaneously located or mucosal ones appear more bluish or greenish [Figures 4 and 5] whereas the deeper intramuscular ones may appear as ill-defined swelling with normal overlying skin. These lesions are soft to the touch and empty on applying pressure as well as on limb elevation. This is compressibility or the sign of emptying [Figure 6a and b]. Sluggish flow and stasis lead to phlebothrombosis, which presents clinically as recurrent pain and tenderness and later may have palpable phleboliths.\(^13\)

**Capillary malformations**

These are also called venular malformations and are recognized as birthmarks or port-wine stains and are common in the trigeminal dermatome distribution.\(^14\) Although in early
stages, they are flat and pink, the CM may evolve into a raised, thickened red to purple plaque as the child matures and eventually may become studded with vascular papules, imparting a cobblestone-like appearance [Figure 7]. Facial lesions in trigeminal nerve distribution can often be a part of the Sturge–Weber Syndrome (SWS) known (improperly) as encephalotrigeminal angiomatosis. SWS is characterized by facial CM and intracranial vascular malformation of the arachnoid and pia mater meninges and presents with intractable seizures, mental retardation, or glaucoma. [2,5]

**Lymphatic malformations**

Based on the size of the lymphatic lumen, LMs (previously termed lymphangiomas) can be divided into microcystic lesions (previously termed lymphangioma circumscriptum) and macrocystic lesions (previously termed cystic hygromas) and a combined form. [15] Lymphatic malformations (LMs) occur most commonly in the head and neck, followed by axilla, chest wall, and extremities. [16] A special scoring system (Cologne Disease Score) based on disfigurement, dysphagia, dysphonia, and dyspnea has been used to predict the outcome of children with LMs. [17] LM present as soft, easily compressible masses [Figure 8] with thin overlying skin that may swell in dependent positions or when venous pressures increase (crying or Valsalva). [18] Bleeding within the cyst or a mixed veno-LM may result in blue discoloration of the overlying skin. Microcystic LMs are soft and noncompressible masses with an overlying area of small vesicles involving the skin or mucosa, which can weep and occasionally cause pain or minor bleeding. [5]

**Arteriovenous malformations**

AVM is a high flow malformation with multiple arteriovenous shunting within a nidus which consists of a capillary network. [19] The most common sites are intracranial followed by extracranial head and neck, extremity, trunk, and visceral. [20] The clinical presentation ranges from an asymptomatic mass to cardiac failure. Superficially located AVMs present as firm mass with warmth, bruit, and thrill. They do not classically empty readily on compression or limb elevation. Bleeding occurs more frequently with AVM than with other vascular malformations, while other presenting symptoms include ulcer, ischemic steal, or skin changes of venous hypertension [Figures 9 and 10].

Table 2 shows the four stages of AVM as proposed by Schobinger. [21]

**Combined vascular malformations**

Some patients have combined malformations which are complex syndromes and are often associated with overgrowth

---

**Figure 3:** Superficial vascular malformation in adult

**Figure 4:** Venous malformation of finger

**Figure 5:** Mucosal venous malformation over tongue

**Figure 6:** (a) Finger venous malformation (b) Compressibility and sign of emptying
of musculoskeletal tissue. They can be classified according to high or low blood flow.[12]

1. Low flow
   a. Klippel–Trenaunay Syndrome (KTS) is a combined capillary, lymphatic and VM in one or more limbs in association with skeletal or soft tissue overgrowth.[22]
      The classical triad includes atypical varicose veins, nevus, and limb overgrowth[10] [Figure 11]. There is the presence of persistent lateral marginal vein of Servelle, which runs from ankle and lateral aspect of leg, thigh and then into the inguinal or gluteal area.[10]
      The main complication of KTS is thrombophlebitis, which is reported in 20%–45% of patients and causes pulmonary embolism in 4%–25% of them[24–26]
   b. Proteus Syndrome is a heterogeneous condition characterized by asymmetric vascular, skeletal, and soft-tissue lesions of varying size.[27]
      Asymmetric body growth and macrodactyly are the classic features with cutaneous or CM spots and small microcystic lymphatic or low flow venous malformations.[28]
   c. Maffucci Syndrome is associated with venous, capillary, and occasionally LMs, with exostosis and enchondromas.[29]
      These enchondromas can lead to deformities or pathological fractures with a long-term possibility of malignant transformation into chondrosarcoma.

2. High flow
   a. Parkes Weber Syndrome is characterized by diffuse reddish pink macule with evenly spreading geometric or blotchy borders. Unlike the Klippel–Trenaunay syndrome, the vascular lesion is a high-flow one with arteriovenous fistulas. Abnormal lateral venous anomalies as well as LMs are uncommon, while musculoskeletal involvement does not usually occur.[30]
      The main complication in the Parkes-Weber syndrome is cardiac failure and cutaneous ischemia.[31]
**Diagnostic Approach to Vascular Malformations**

**Hematologic evaluation**

Coagulation disorders occur at a high frequency in patients with CVMs and are associated with potentially severe thromboembolic events and hemorrhagic complications. Thromboembolic and hemorrhagic complications in CVM patients have been reported following sclerotherapy, surgery, trauma, prolonged immobilization, hormonal changes including pregnancy and menstruation, and sepsis.

Localized intravascular coagulopathy (LIC) occurs due to stasis within these vessels with formation of thrombin and subsequent conversion of fibrinogen to fibrin, which is followed by fibrinolysis and evidence of fibrin degradation products like D-dimer.

LIC is of important clinical concern due to the potential for more serious thromboembolic events, including superficial thrombosis, deep venous thrombosis, or pulmonary embolism as well as thrombohemorrhagic disseminated intravascular coagulation (DIC) with life-threatening hemorrhage, which can occur during or following surgical resection or sclerotherapy. This is essentially different from the Kasabach-Merritt Syndrome, which is a distinct clinical entity characterized by DIC and profound thrombocytopenia (<50,000/ml) frequently associated with vascular tumors including hemangiomas. By contrast, the platelet count in LIC is minimally diminished (100,000–150,000/ml range).

Conversion of LIC to DIC can be detected early by an increased prothrombin time as well as reduction in platelet counts. Extensive CVM with large surface area, muscle involvement, and/or palpable phleboliths are strong predicting criteria for coagulation disorders associated with CVM. Assessment of the coagulation profile and D-dimer levels is indicated in patients with extensive CVMs.

LIC as characterized by elevated D-dimer levels has been observed in approximately 40% of patients with CVMs. Patients with severe LIC would present with highly elevated D-dimer levels associated with low fibrinogen levels. Anticoagulation with Low-molecular-weight heparin (LMWH) can be used to treat the pain caused by LIC and to prevent decompensation of severe LIC to DIC.

In summary, patients with extensive VMs or high-risk lesions in particular should undergo the laboratory tests as enlisted in Table 3.

**Ultrasound and Doppler**

The diagnosis of a CVM can often be made with a careful history and physical examination. Noninvasive imaging is used to confirm the diagnosis. Ultrasonography (US) and magnetic resonance imaging (MRI) are the most widely used modalities of choice.

For US to be a useful modality, it must include gray-scale, color Doppler, and spectral Doppler tracings to evaluate vascularity and determine types of vessels present. VMs are compressible with the typical multispacial, multicystic, and/or partially solid heterogeneous echotexture (98%) and can be hypechoic (82%), hyperechoic (10%), or isoechoic (8%) with respect to surrounding structures. Although phleboliths are classic of VM and noted on plain X-ray, they are rarely detected on US.

On color Doppler examination, monophasic waveforms with low flow is noted in VM, whereas continuous high flow is noted in AVM and no flow noted in large hypechoic cysts in LM. The presence of pulsatile triphasic flow of nearby arteries in a VM should not be confused with AVM.

**Magnetic resonance imaging**

MRI is helpful to further characterize sonographic findings and determine the extent of larger lesions for planning medical, surgical, or interventional procedures.

---

**Table 3: Hematologic investigations for congenital vascular malformations**

| Routine | Full blood count including hemoglobin levels and platelet count D-dimer - quantitative assay |
|---------|------------------------------------------------------------------------------------------|
| If LIC suspected | Fibrinogen PT, APTT Thrombophilia screening |

APTT: Activated partial thromboplastin time, PT: Prothrombin time, LIC: Localized intravascular coagulopathy

**Table 4: Yakes classification of arteriovenous malformations**

| Type | Manifestation |
|------|---------------|
| Type 1 | Direct AV fistula |
| Type 2A | Typical AVM nidus (multiple inflow arteries leading to nidus and vein outflow) |
| Type 2B | Typical nidus with aneurysmal venous drainage (dominant venous outflow) |
| Type 3A | Aneurysmal vein wall acts as the nidus |
| Type 3B | Multiple vein wall acts as the nidus |
| Type 4 | Infiltrative AVM permeating a tissue |

AVM: Arteriovenous malformations, AV: Arteriovenous

---

Figure 11: Klippel–Trenaunay Syndrome on the right lower limb. Note the large lateral marginal vein
interventional, and/or surgical therapy. Recommended sequences are T1- (pre- and post-contrast) and T2-weighted images, with fat saturation. Typically, images may have an intermediate signal intensity in T1 and a hyperintense signal in T2 in relation to its content or the presence of hemorrhage or thrombosis. A gadolinium injection usually shows a diffuse enhancement of serpentine venous channels, unlike LMs, which generally do not have this kind of enhancement [Figure 13]. With T2-weighted images, phleboliths appear as focal areas of hypointense signal. In macrocystic LM, hyperintense signal in T2 images and low-intensity signal in T1 images, with postcontrast enhancement of the septa is noted whereas, microcystic lesions generally appear as homogeneous hyperintense signals in T2 images. Numerous hypolucent arterial flow voids without an obvious mass are the hallmark of AVM by MRI.

**Computed tomography scan**

Computed tomography (CT) is of little use in imaging the VM, except in the examination of bony VM. However, AVMs appear as a tortuous collection of vessels with one or more enlarged feeding arteries [Figure 14]. Intravascular contrast material shunts rapidly into enlarged draining veins which may sometimes be aneurysmal. In an asymptomatic AVM, there should be no associated soft-tissue enhancement. The presence of soft-tissue enhancement should raise the possibility of a tumor, such as a sarcoma. Due to the risk of radiation with CT scan, an MRI imaging with a time-resolved MR angiography can be a very useful alternative.

**Digital subtraction angiography**

Catheter-based angiographic evaluation of an AVM and its behavior allows strategic decision making with regard to the approach to embolization. The classic digital subtraction angiography (DSA) appearance is tortuous vessels with enlarged feeding arteries, which rapidly shunt into dilated draining veins by way of a nidus [Figure 15]. Depending on the angiography findings, four types of AVM architecture have been described by Cho et al. Another classification utilized for peripheral AVM is by Yakes [Table 4].

DSA is absolutely required before treatment to assess flow rate, visualize anatomy of the nidus in greater detail than magnetic...
resonance angiography, and identify vessels required for distal circulation.\cite{32}

**TREATMENT**

**General principles for congenital vascular malformation**

The multidisciplinary team approach is recommended with the aim of proper selection or combination of surgical, nonsurgical, or endovascular treatment methods.\cite{32,53} Some guiding general principles are as follows:\cite{32}

1. Not every CVM is amenable to treatment
2. Not every CVM needs to be treated aggressively
3. An overzealous approach sometimes does more harm than good\cite{54}
4. Observation sometimes remains the best approach yet until figuring out the exact nature of the lesion
5. When the benefit of treatment outweighs the risk of complications and morbidity, less risky treatment options should be first-line therapy
6. In contrast to the treatment of AVMs, all VM lesions can be treated using a less aggressive approach
7. It is usually safe to delay treatment until the child reaches to the age of two or more years before beginning diagnostic procedures and treatment\cite{55}
8. However, for the VM lesion at a life- or limb-threatening anatomic location, an earlier treatment approach is preferred over a more conservative one
9. In limb- and life-threatening situations, sacrificing limb over life may be necessary
10. Complicated patients may be referred to an experienced center in time.

**Conservative management**

Proper skin care, wound dressings, compression bandages, or stockings as well as orthopedic footwear to correct deformities are the foremost means of conservative treatment.\cite{32} Extensive, multifocal, infiltrating, and painful CVMs should be treated with weight-adjusted dose (100 U/kg/d) of LMWH.\cite{40} Certain indications for treatment of CVM are enlisted in Tables 5 and 6.

**Sclerotherapy**

Sclerotherapy is the mainstay of treatment of CVMs. The goal of sclerotherapy is to obliterate the vascular channels by causing damage to the endothelium with subsequent inflammation and fibrosis. Sclerotherapy can be performed under direct vision, Doppler guidance, or fluoroscopy control [Figure 16a and b]. Among all the sclerosing agents, absolute ethanol is the most effective one with the lowest recurrence rate but also with the most serious local and systemic side effects.\cite{56,57} For large lesions, multiple treatments are often necessary.\cite{58} Some publications have shown a range of 1–12 sessions for low flow VM to achieve clinical benefit.\cite{58,61}

Ethanol is effective for treatment of large, extensive VMs but should be used with caution as it can damage nerves, cause skin necrosis, and induce systemic toxicity including pulmonary vasospasm and cardiovascular collapse.\cite{32} It may also not be suitable for use in mucosal surface or thin skin. The volume of ethanol (60-99% concentration) that can be injected safely is accepted to be 0.15 ml/kg over 10 min (no Swan-Ganz monitoring is necessary at this dose).\cite{62} Ethanol sclerotherapy for VM as well as AVM should be performed under sedation or general anesthesia, as it is very painful. A proximal tourniquet is usually not required. After confirmation of blood flow after needle or scalp vein puncture, contrast study is performed till draining veins are noted. The dosage of absolute ethanol is approximately ¼–2/3 of the amount of the contrast used.\cite{58} Perioperative useful measures include dexamethasone to reduce tissue edema as well as 5% dextrose with sodium bicarbonate for urine alkalinization to prevent acute renal failure by hemoglobinuria.\cite{40,58}

Other agents used include polidocanol or sodium tetradecyl sulfate as foam sclerosants (0.5–3% concentration) and bleomycin. Bleomycin is useful in treating VM and LM in
sensitive locations such as the orbit and airway, as it leads to minimal postprocedural swelling.\textsuperscript{[32]} Macrocystic LM respond well to sclerotherapy with bleomycin or doxycycline whereas microcystic fare poorly.\textsuperscript{[48]} The most common postprocedure side effects of sclerotherapy are pain and inflammation. In some extensive CVM involving the head and neck, tracheostomy might be required during the treatment sessions to secure airway.\textsuperscript{[5,58]} A large retrospective study found slow appearance of drainage vein on initial direct puncture venography, well-defined margins on MRI, and female gender to be statistically significant, independent factors predictive of a good response to ethanol sclerotherapy in patients with VM.\textsuperscript{[63]}

**Lasers**

Endovenous thermal ablation under ultrasound guidance by diode LASER may have an adjunctive role in the therapy of large truncular VMs as well as large soft-tissue phlebectasias.\textsuperscript{[32,64]} Similar use of Nd: YAG laser photocoagulation has been published for mucosal or superficial malformations.\textsuperscript{[65]}

**Surgery**

Open surgical excision combined with the endovascular therapy (embolotherapy/sclerotherapy) is the most effective means to control extratruncular VM lesions.\textsuperscript{[32,66]}

The proximal surgical ligation of feeding arteries without resection (and endovascular coiling of feeder arteries) is doomed to failure and must be avoided as it does not cure the nidus which allows the AVM to grow. Furthermore, these procedures make a subsequent access and therapeutic embolization difficult.

Simple well-localized VM can be excised surgically with good results [Figure 17a-d]. However, controversies exist in the role of debulking surgeries for extensive VM as they are associated with significant blood loss and there is a tendency of these extratruncular VM to regrow.\textsuperscript{[67]} Predictors of significant blood loss during surgery include high flow AVM, inability to use tourniquet, debulking surgery, and low platelet count.\textsuperscript{[68]} However, in selected patients, total surgical excision for well-defined and localized VM as well as partial excision for debulking of extensive VM has been noted to give nearly 75% clinical and symptomatic improvement [Figure 18a-c]. Lymphatic malformations tended to be nonoperable, and when operable, they are unlikely to be completely excised.\textsuperscript{[69]}

For AVM, a combined approach with preoperative embolotherapy/sclerotherapy is often implemented for surgically accessible lesions whenever feasible to reduce surgical morbidity as surgical excision offers the best opportunity for “cure.”\textsuperscript{[70]} Surgery ideally performed between 24 and 48 h of embolization is aimed at removal of the malformation with a margin of 5 mm to 1 cm, with immediate reconstruction whenever possible.\textsuperscript{[71]}

Although the first-line management of patients with KTS continues to be nonoperative, those patients with patent deep veins can be considered for excision of symptomatic varicose veins and VMs with improved clinical outcomes.\textsuperscript{[72]}

**Embolotherapy**

The goal of AVM embolization (or embolotherapy) is to obliterate the nidus while simultaneously minimizing nontarget embolization.\textsuperscript{[52,73]} Embolization may be performed by different approaches: direct puncture of the nidus or embolization performed by way of a transarterial or transvenous method.\textsuperscript{[47,74]} In treating AVMs endovascularly, superselective catheter placement through the arterial route in the nidus is essential; however, if not feasible to enter the nidus, then direct percutaneous puncture techniques should be employed.\textsuperscript{[47]}

Selection of the appropriate agent for embolotherapy is as essential as correct patient selection. Various embolic materials available include ethanol, N-butyl cyanoacrylate (NBCA), polyvinyl alcohol particles, ethylene vinyl alcohol copolymer (Onyx), and endovascular coils and vascular plugs.\textsuperscript{[32]}

---

**Figure 17:** (a) Venous malformation in preauricular area prepared for surgical excision (b) Complete excision of preauricular venous malformation done without significant bleeding (c) After complete excision (d) Final picture after sutures

**Figure 18:** (a) Large venous malformation over forearm and hand (b) Debulking surgery performed under tourniquet control (c) Two years after debulking surgery
Following successful placement in the nidus, ethanol embolization is performed in small 1-3cc aliquots at a total dose of 0.15 ml/kg. Ethanol can be mixed with iodized oil (Lipiodol) if visualization is desired. If higher dose is expected (up to 0.5 ml/kg), a Swan Ganz catheter is placed for pulmonary arterial pressure (PAP) measurements.\[51,52\] If PAP rises above 25 mmHg systolic, then treatment with nitroglycerin infusion at 1 µg/kg/min is required.\[75\] If superselective placement in an AVM nidus is not possible, then the use of ethanol must be avoided. Postprocedure, monitoring and precautions are taken as mentioned earlier.

NBCA or glue is a liquid-casting adhesive agent generally considered to be safer than ethanol and needs to be diluted with lipiodol (to make it radiopaque) at a ratio of 1:2 or 1:3 and has an expected polymerization time of 1–4 s.\[52\] It polymerizes quickly and irreversibly when exposed to anions in blood and is effective even in the presence of coagulopathy. Hence, it is used carefully with polypropylene catheters, using 5% dextrose as a flushing solution.\[76\] It requires special skill to prevent nontarget polymerization or reflux in feeding artery as well as adherence of catheter. The classic appearance of radio-opaque cast of glue-lipiodol is noted within the nidus [Figure 19].

Onyx (6-8%) has the advantage of ease of use and slower polymerization with adequate distal penetration from intra-arterial access [Figure 20a and b]. It is mixed with dimethyl sulfoxide (DMSO) and is injected slowly; the maximum injection rate is 0.1 ml/min to avoid vasospasm caused by DMSO.\[77\] The main advantage of Onyx over NBCA is the slow flow and longer casting time, giving the operator greater control over administration. The disadvantage is longer treatment times and greater expense, with possibility of skin tattooing in superficial lesions.\[52\]

Endovascular coils and plugs have a limited role in peripheral AVM although they have been successfully used in pulmonary and renal AVM and may occasionally be indicated for dominant outflow vein occlusion.\[78\] Often a combination of different agents is required for successful embolization.

**Follow-up**

Close postoperative observation with expected management of local recurrence is required. On early follow-up, one must watch for complications as well as DVT. After completion of repeat treatment sessions, a 6 monthly or annual Doppler or MRI is recommended to assess the effectiveness of treatment and detect persistence or late recurrence.\[32\] If treated appropriately, most patients will experience at least symptomatic improvement after endovascular therapy and possible cure after surgery.

**Conclusion**

Vascular anomalies remain one of the most complex and ill-understood diseases treated by a vascular surgeon. A proper knowledge of classification and correct diagnosis helps in deciding and delivering appropriate treatment. Treatment of vascular anomalies is challenging and often involves various therapeutic options. Multidisciplinary approach with full integration of open surgical and endovascular therapy has become the mainstay of treatment in the contemporary management of CVMs.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

---

**Figure 19:** Radio-opaque cast of glue + lipiodol in arteriovenous malformation nidus of the foot

**Figure 20:** (a) Digital subtraction angiography shows arteriovenous malformation nidus of the ear (b) Successful Onyx embolization of arteriovenous malformation nidus using Onyx and microcatheter. The feeding artery remains patent.
Conflicts of interest

There are no conflicts of interest.

References

1. Enjolras O. Classification and management of the various superficial vascular anomalies: Hemangiomas and vascular malformations. J Dermatol 1997;24:701-10.
2. Colletti G, Valassina D, Bertossi D, Melchiorre F, Vercellio G, Brusati R, et al. Contemporary management of vascular malformations. J Oral Maxillofac Surg 2014;72:510-28.
3. Chiller KG, Passaro D, Frieden IJ. Hemangiomas of infancy: Clinical characteristics, morphologic subtypes, and their relationship to race, ethnicity, and sex. Arch Dermatol 2002;138:1567-76.
4. Fimm MC, Glowacki J, Mulliken JB. Congenital vascular lesions: Clinical application of a new classification. J Pediatr Surg 1983;18:894-900.
5. Richter GT, Friedman AB. Hemangiomas and vascular malformations: Current theory and management. Int J Pediatr 2012;2012:645678.
6. Shayan YR, Prendiville JS, Goldman RD. Use of propranolol in treating hemangiomas. Can Fam Physician 2011;57:302-3.
7. Oak SN, Viswanath N. Management of hemangiomas in children. Indian J Dermatol Venerol Leprol 2006;72:1-4.
8. Cox JA, Bartlett E, Lee EI. Vascular malformations: A review. Semin Plast Surg 2014;28:58-63.
9. Lee BB, Laredo J, Lee TS, Huh S, Neville R. Terminology and classification of congenital vascular malformations. Phlebology 2007;22:249-52.
10. Lee BB, Baumgartner I, Berlien P, Bianchini G, Burrows P, Gloviczki P, et al. Diagnosis and treatment of venous malformations. Consensus document of the International Union of Phlebology (IUP): Updated 2013. Int Angiol 2013;34:97-149.
11. Wassef M, Blei F, Adams D, Alomari A, Basela B, Berenstein A, et al. Vascular anomalies classification: Recommendations from the international society for the study of vascular anomalies. Pediatrics 2015;136:e203-14.
12. Redondo P. Vascular malformations (I). Concept, classification, pathogenesis and clinical features. Actas Dermosifiliogr 2007;98:141-58.
13. Ethunandan M, Mellor TK. Haemangiomas and vascular malformations of the maxillofacial region – A review. Br J Oral Maxillofac Surg 2006;44:263-72.
14. Syed MN. Vascular lesions of head and neck: A literature review. Indian J Dent Sci 2016;8:176-82.
15. Khunger N. Lymphatic malformations: Current status. J Cutan Aesthet Surg 2010;3:137-8.
16. Lee BB, Kim YW, Seo JM, Hwang JH, Do YS, Kim DI, et al. Current concepts in lymphatic malformations. Vasc Endovascular Surg 2005;39:67-81.
17. Wittekind C, Michel O, Streppel M, Roth B, Quante G, Beutner D, et al. Lymphatic malformations of the head and neck: Introduction of a disease score for children, cologne disease score (CDS). Int J Pediatr Otorhinolaryngol 2006;70:1205-12.
18. Lowe LH, Marchant TC, Rovid DC, Scherbel AJ. Vascular malformations: Classification and terminology the radiologist needs to know. Semin Roentgenol 2012;47:106-17.
19. Marler JJ, Mulliken JB. Current management of hemangiomas and vascular malformations. Clin Plast Surg 2005;32:99-116, ix.
20. Mattassi R. Loose DA, Vaghi M. Hemangiomas and Vascular Malformations: An Atlas of Diagnosis and Treatment. Italy: Springer Verlag Italia; 2003.
21. Shohingber R. In: Proceedings of International Society for the Study of Vascular Anomalies Congress, Rome, Italy; 23-26 June, 1996.
22. Servelle M, Klippel and trenaunay’s syndrome 768 operated cases. Ann Surg 1985;201:365-73.
23. Oduber CE, Young-Afat DA, van der Wal AC, van Steensel MA, Hennekam RC, van der Horst CM, et al. The persistent embryonic vein in klippel-trenaunay syndrome. Vasc Med 2013;18:185-91.
24. Baskerville PA, Ackroyd JS, Lea Thomas M, Browse NL. The klippel-trenaunay syndrome: Clinical, radiological and haemodynamic features and management. Br J Surg 1985;72:232-6.
25. Gloviczki P, Stanson AW, Stickler GB, Johnson CM, Toomey BJ, Meland NB, et al. Klippel-trenaunay syndrome: The risks and benefits of vascular interventions. Surgery 1991;110:469-79.
26. Samuel M, Spitz L. Klippel-trenaunay syndrome: Clinical features, complications and management in children. Br J Surg 1995;82:757-61.
27. Wiedemann HR, Burgio GR, Aldenhoff P, Kunze J, Kaufmann HJ, Schirg E, et al. The proteus syndrome. Partial giantism of the hands and/or feet, nevi, hemihypertrophy, subcutaneous tumors, macrocephaly or other skull anomalies and possible accelerated growth and visceral affections. Eur J Pediatr 1983;140:5-12.
28. Hotamisiligil GS. Proteus syndrome and hamartoses with overgrowth. Dysmorphol Clin Genet 1990;4:87-102.
29. Kaplan RP, Wang JT, Amron DM, Kaplan L. Maffucci’s syndrome: Two case reports with a literature review. J Am Acad Dermatol 1993;29:894-9.
30. Ziyeh S, Spreer J, Rössler J, Strecker R, Hochmuth A, Schumacher M, et al. Parkes weber or klippel-trenaunay syndrome? Non-invasive diagnosis with MR projection angiography. Eur Radiol 2004;14:2025-9.
31. Cohen MM Jr. Vascular anomalies: Classification, angiogenesis, hemangiomas, and vascular malformations. Am J Med Genet 2002;108:265-74.
32. Lee BB, Bergan J, Gloviczki P, Laredo J, Loose DA, Mattassi R, et al. Diagnosis and treatment of venous malformations. Consensus document of the international union of phlebology (IUP)-2009. Int Angiol 2009;28:434-51.
33. Mazoyer E, Enjolras O, Bisdorff A, Perdu J, Wassef M, Drouet L, et al. Coagulation disorders in patients with venous malformation of the limbs and trunk: A case series of 118 patients. Arch Dermatol 2008;144:861-7.
34. Oduber CE, Gerdes VE, van der Horst CM, Bresser P. Vascular malformations as underlying cause of chronic thromboembolism and pulmonary hypertension. J Plast Reconstr Aesthet Surg 2009;62:684-9.
35. Mason K, Neufeld EJ, Karian VE, Zurakowski D, Koka BV, Burrows PE, et al. Coagulation abnormalities in pediatric and adult patients after sclerotherapy or embolization of vascular anomalies. AJR Am J Roentgenol 2001;177:1359-63.
36. Martin LK, Russell S, Wargon O. Chronic localized intravascular coagulation complicating multifocal venous malformations. Australas J Dermatol 2009;50:276-80.
37. Levi M. Current understanding of disseminated intravascular coagulation. Br J Haematol 2004;124:567-76.
38. Mazoyer E, Enjolras O, Laurian C, Houdart E, Drouet L. Coagulation abnormalities associated with extensive venous malformations of the limbs: Differentiation from kasabach-merritt syndrome. Clin Lab Haematol 2002;24:243-51.
39. Dompmartin A, Ballieux F, Thibon P, Lequerrec A, Hermans C, Clapuyt P, et al. Elevated D-dimer level in the differential diagnosis of venous malformations. Arch Dermatol 2009;145:1239-44.
40. Dompmartin A, Acher A, Thibon P, Tourbach S, Hermans C, Deneyes V, et al. Association of localized intravascular coagulopathy with venous malformations. Arch Dermatol 2008;144:873-7.
41. Dubois J, Garel L. Imaging and therapeutic approach of hemangiomas and vascular malformations in the pediatric age group. Pediatr Radiol 1999;29:879-93.
42. Trop I, Dubois J, Guibaud L, Grignon A, Patriquin H, McCuaig C, et al. Soft-tissue venous malformations in pediatric and young adult patients: Diagnosis with doppler US. Radiology 1999;212:841-5.
43. Olivieri B, White CL, Restrepo R, McKoon B, Karakas SP, Lee EY, et al. Low-flow vascular malformation pitfalls: From clinical examination to practical imaging evaluation – Part 2, venous malformation mimickers. AJR Am J Roentgenol 2016;206:952-62.
44. Siere S, Teplisky D, Lipsch J. Vascular malformations: An update on imaging and management. Arch Argent Pediatr 2016;114:167-76.
45. Dubois J, Alison M. Vascular anomalies: What a radiologist needs to know. Pediatr Radiol 2010;40:895-905.
46. Donnelly LF, Adams DM, Bisset GS 3rd. Vascular malformations and hemangiomas: A practical approach in a multidisciplinary clinic. AJR Am J Roentgenol 2000;174:97-108.
47. Dunham GM, Inghram CR, Maki JH, Vaidya SS. Finding the nidus: Detection and workup of non-central nervous system arteriovenous malformations. Radiographics 2016;36:891-903.
48. Mulligan PR, Prajapati HJ, Martin LG, Patel TH. Vascular anomalies:
