*Review Article*

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# Current Guidelines for Vascular Anomalies Management

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**Abstract**

Vascular anomalies are a heterogeneous group of rare and complex disease that can occur anywhere in the human body. These anomalies pose a major challenge in diagnosis and treatment because of their multiple clinical presentations and anatomic variations. The treatment of vascular anomalies requires a multidisciplinary approach involving specialists from different medical fields such as radiology, interventional radiology, plastic surgery, vascular surgery, dermatology and hematology. The key to effective treatment lies in the accurate diagnosis and classification of these lesions, which can be achieved through a combination of clinical evaluation, imaging studies, and histopathologic analysis. Treatment modalities for vascular malformations include a wide range of options, including conservative observation, minimally invasive techniques, and surgical intervention. Noninvasive methods such as compression garments and sclerotherapy have been shown to be effective in vascular anomalies.

**Keywords:** Guidelines; Vascular anomalies; Management.

## INTRODUCTION

Vascular anomalies refer to a diverse group of congenital anomalies affecting the blood vessels, ranging from simple to complex manifestations. These anomalies may involve either abnormal growth or development of blood vessels, resulting in a wide range of clinical manifestations. Vascular anomalies are broadly classified into two main groups: vascular tumors and vascular malformations. Vascular tumors, such as infantile hemangiomas, involve excessive growth of blood vessels that typically undergo a phase of rapid proliferation followed by slow regression. Vascular malformations, on the other hand, are structural abnormalities that are present from birth and involve defects in blood vessel development that do not spontaneously regress. Examples include venous malformations, lymphatic malformations, and arteriovenous malformations. Accurate diagnosis and appropriate management of vascular anomalies are critical because they can vary widely in their clinical course, potential, and impact.

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## CLASSIFICATION

The newer classification updated by ISSVA in 2018 is as follows in Table 1

**Table 1:** ISSVA classification for vascular anomalies (Approved at the 20th ISSVA Workshop, Melbourne, April 2014, last revision May 2018)[1]

| Vascular anomalies | | | | |
|---|---|---|---|---|
| **Vascular tumors** | **Vascular malformations** | | | |
| | **Simple** | **Combined** | **of major named vessels (channel type or truncal vascular malformations)** | **associated with other anomalies** |
| - Benign<br>- Locally aggressive/ borderline<br>- Malignant | - Capillary malformation<br>- Lymphatic malformation<br>- Venous malformation<br>- Arteriovenous malformation<br>- Arteriovenous fistula | CVM<br>CLM<br>LVM<br>CLVM<br>CAVM<br>CLAVM<br>CVAVM<br>CLVAVM | - Affect lymphatics veins arteries<br>- Anomalies of Origin, course, number, length, diameter (aplasia, hypoplasia, stenosis, ectasia / aneurysm), valve communication (AVF), persistence of embryonal vessel | - Klippel-Trenaunay syndrome<br>- Parkes Weber syndrome<br>- Servelle-Martorell syndrome<br>- Sturge-Weber syndrome<br>- Maffucci syndrome<br>- CLOVES syndrome<br>- Proteus syndrome<br>- Bannayan-Riley-Ruvalcaba syndrome<br>- CLAPO syndrome |

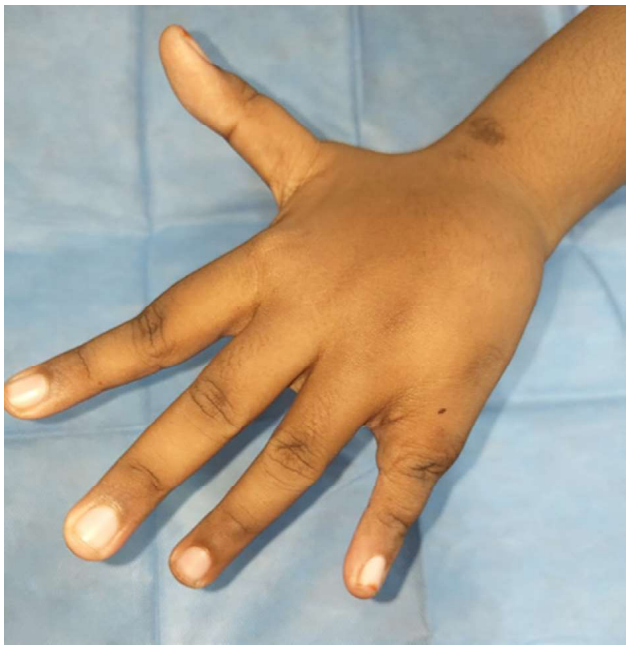

**Fig. 1:** Lymphovenous malformation of hand

### *Clinical Evaluation*

The diagnostic algorithm includes a thorough history and a detailed physical examination. The history is to know whether it is present since birth and the rate of growth. The physical examination should include careful assessment (inspection, palpation, auscultation) of the lesion and arterial, venous and lymphatic systems. It includesan assessment of blood pressure and peripheral pulses on the affected side and the contralateral limbs as well as other findings like dilated vessels/ varicosities, thrombophlebitis, local temperature, edema, skin changes like pigmentation, induration, ulcerations and other changes of chronic venous insufficiency (CVI). Clinical examination of the involved limb should include an assessment of the limb size, volume, symmetry, co-existing soft tissue, or bony hypertrophy or atrophy.

### *Diagnostic Evaluation*

Diagnostic algorithms for vascular anomalies should start with an ultrasound investigation first, followed by non-invasive MR or CT scans, and finally invasive investigations like angiographies. Additional investigations include nuclear medicine imaging such as radionuclide lymphoscintigraphy, whole body blood pool scintigraphy (WBBPS), and transarterial lung perfusion scintigraphy (TLPS)

## NON-INVASIVE DIAGNOSTIC EVALUATION

### *Duplex Ultrasonography (DUS)*

Ultrasound is the most important diagnostic imaging modality for vascular anomalies. The advantages of ultrasound are its widespread use, low cost, and lack of radiation exposure, which make it the most important tool for the study of vascular anomalies.[2,3,4]

Limitations include the inability to define the extent of lesions that are not in the extremities and the need for MRI findings for correlation in deep seated lesions. It is also limited when air is present (e.g., in the lungs) or when abnormalities are localised within bone. Bony abnormalities can be studied with other imaging modalities, such as CT or transcranial devices, if they are located within the skull. Limitations in the hemodynamic evaluation of occlusive venous status by ultrasound can be avoided by using intravascular ultrasound (IVUS).

### *MRI*

It is the procedure of choice after ultrasonography. It has better spatial resolution and a wider field of view than ultrasound. It also provides the ability to visualize blood flow and tissue characteristics without ionizing radiation.[5,6] When examining vascular abnormalities, high-power devices (at least 15 Tesla) should be used to achieve better contrast and spatial resolution. The disadvantage of MRI is that it is lengthy, noisy, and potentially frightening for children, especially in younger age groups where consistent sedation is required for MRI. A typical CVM imaging protocol consists of T1-weighted spin echo (SE) or fast spin echo (FSE) images of the lesion, generally with fat suppression. Images obtained after gadolinium injection are useful to distinguish between LMs and VMs that have similar images on normal and angiographic imaging. SE Sequences can also identify signal gaps representing arterial feeders. T2-weighted images (FSE with fat suppression or, alternatively, short tau inversion recovery STIR images) in at least two planes were found to be most sensitive and specific for detecting the extent and depth of the lesion. Lesion over a fat, muscle, and bone background with low signal intensity; these sequences may also show the content of the malformation. Using bright blood gradient recall echo sequences (GRE) to identify high flow vessels as rounded and hyperintense signal gaps.

### *CT*

CT is an alternative to MRI in patients with respiratory or cardiac failure in whom sedation with MRI is contraindicated,[6] in vascular abnormalities of the bowel and lungs, and in patients with embolic spirals or metal clips. CT is useful in changes in bone architecture and identifies phleboliths or other dystrophic calcifications. Adverse effects include exposure to ionizing radiation and the need to use a contrast agent to visualize the vessels.

On CT, VMs often appear as hypodense or heterogeneous lesions that slowly enlarge from the periphery after injection of contrast medium. CT Venography is of unique value in evaluating obstructed anomalous, atretic, or absent veins and other truncal abnormalities of large veins in the chest, abdomen, or pelvis. CT accurately identifies underlying pathology, confirms venous obstruction or extrinsic compression, describes anatomic variations, and the extent of venous thrombosis. LMs show fluid-filled masses with low attenuation, occasionally with fluid content and peripheral contrast enhancement of the wall. Contrast-enhanced CT of AVMs with bolus-tracking technique (CT-angiography, CTA) to obtain optimal study of arterial vessels shows numerous enlarged feeding arteries with rapid contrast shunt into enlarged draining veins without significant intervening tissue enhancement that would normally be seen in a normal capillary network. Contrast-enhanced CT of AVMs is significantly more informative than other vascular malformations, providing a unique three-dimensional data set for accurate mapping and measurement of arterial, nidal, and venous structures and assessment of flow patterns for interventional radiologic or surgical planning, especially given the many post-processing options.

VMs are heterogeneous lesions that slowly enlarge from the periphery after contrast injection. In AVMs, the feeding artery, draining veins, and nidus are well defined.

CT with intravenous contrast enhancement has been used for differential diagnosis of hemangiomas and VMs.

CTA provides much better anatomic information and sometimes shows arterial and venous anatomy in excellent detail, but is inferior in all aspects to the newer technique of CE-MRI.

### *Invasive Diagnostic Evaluation*

- Ascending, descending, and/or segmental venography/phlebography
- Standard and/or selective arteriography
- Percutaneous direct puncture angiography arteriography, phlebography, varicography, lymphography

The basic diagnosis of CVMs is usually adequate with the right combination of non to minimally invasive tests. "Invasive" tests are rarely needed to make the diagnosis of VM and can be deferred until surgery is needed. They are required for treatment planning, either surgical or endovascular. However,

invasive tests may be required for diagnosis if non minimally invasive tests (e.g., CT and/or MRI) do not confirm the diagnosis or reveal important diagnostic details that are important for treatment options. These tests are required before treatment with embolotherapy. Direct puncture phlebography is also very useful to identify a large draining vein in extratruncal lesions. These veins can be treated in advance to allow more effective therapy with less risk of recurrence, with subsequent embolotherapy or sclerotherapy.

There is no role for diagnostic angiography in the diagnosis or follow-up of low-flow vascular malformations. Angiography used in AVM to findr define the lesion and plan treatment.

### *Phlebography*

Phlebography determines hemodynamic characteristics of the lesion. It also allows the classification based on its anatomy. Based on the appearance of the VMs and the draining venous system during phlebography all VMs can be classified into four distinct groups[7]:

***Type I*** (isolated VMs without phlebographically appreciable venous drainage),

***Type II and Type III*** VMs (demonstrate normal sized and enlarged venous drainage, respectively)

***Type IV*** VMs (characterized by essentially ectatic dysplastic veins)

Ascending phlebography is rarely needed to diagnose a vascular malformation. The role of ascending phlebography is limited to the diagnosis of truncular venous disease and especially obstructive pathology. According to non-invasive studies, it is important to evaluate the patency of deep veins in dilated, superficial veins and hypoplasia or aplasia of deep veins.

The main advantage of this method is that imaging can be done in an orthostatic position. Descending phlebography is prescribed for avalvulia and in pelvic, sciatic, and visceral malformations. Direct puncture venography has been implicated in the treatment of extratruncular VM lesions. With this method, it is possible to visualize the outflow of anomaly, and four types can be distinguished according to the veins.

### *Whole body blood pool scintigraphy (WBBPS)*

WHHIPS using Te99 is a very useful tool for detecting vascular malformations. The advantage is the ability to examine entire anatomical structures with a single examination. It provides quantitative information on the hemostasis of the lesion and allows evaluation of treatment results.[8,9,10] It is an excellent tool for routine monitoring and treatment evaluation to assess treatment progress and the natural course of the VM lesion. WHIPS is also an excellent choice for evaluating AVM as well. However, it is more useful for whole-body occult CVM lesion screening as well as for qualitative analysis of AVM lesion during multi-session treatment as a cost-effective procedure. It is an excellent tool for routine monitoring of treatment progress and its natural course even when TLPS is not possible/available.

### *Transarterial lung perfusion scintigraphy (TLPS)*

TLPS finds the degree of AV shunting by the AVM lesion within an extremity. TLPS[11] used especially to assess a micro-AV shunting lesion, which is often difficult with conventional techniques. Micro-AVMs frequently exist in the combined form of CVM. TLPS is of importance in its detection.

### *Radionuclide Lymphoscintigraphy (LSG)*

LSG is a functional study that complements the anatomical information provided by lymphangiography. It is performed by injection of 99mTc-labeled human serum albumin or 99mTc-labeled Sulphur Colloid subcutaneously into the first and second web-space of the toes or fingers, is the test of choice to confirm or exclude lymph vessel pathology as the cause of chronic limb swelling.[12,13]

It assess the flow of colloid from the injection site, transition time to the knee, groins or axilla, absence or presence of major lymphatic collectors, number and size of vessels and nodes, the presence of collateral, reflux and symmetric activity with the opposite side.

SG represents the main examination to evaluate the lymphdynamics of the limbs. It is recorded in rest, after exercise, and one hour of daily activity. It is possible to detect the presence of deep and superficial lymphatic vessels and the presence or absence of reflux.

LSG is used to rule out lymphatic dysfunction due to the presence of a truncular LM known as primary lymphedema, which often exists with the VM. LSG is considered as the gold standard for lymphatic function evaluation as it can clearly indicate lymphatic function.

### *Laboratory Tests*

Coagulation disorders are common in patients with extensive VM and can lead to potentially serious thromboembolic events and bleeding complications.[14] Extensive circulatory disorders are often accompanied by local intravascular coagulopathy (LIC). Evaluation of the coagulation profile and D-dimer level is indicated in patients with extensive VM. D-dimer (a breakdown product of cross-linked fibrin), measured by a rapid enzyme linked immunofluorescence assay, is increasingly used in the evaluation of patients with VM and is considered the biochemical gold standard to rule out thrombophlebitis or a thromboembolic episode. Procedures D-dimer can detect signs of consumptive coagulopathy common in VMs.

***Patients with extensive VMs or high risk lesions in particular should undergo the following laboratory tests:***

- Full blood count including hemoglobin levels and platelet count
- D-dimer-quantitative assay
- Fibrinogen
- PT, APTT
- Thrombophilia screening

### *Histology*

Biopsy should be reserved to make an accurate diagnosis and is mostly required when the lesion is suspected to be a tumor. It differentiates between AVMs, NICH and vascular sarcomas. Biopsy may also be required to differentiate between GVM and BRBNS, GVM are lined by cuboidal glomus cells that stain positive for smooth muscle actin and myosin and is histologically differentiable from BRBNS.[15,16]

### *Endoscopic Evaluation*

Endoscopic examinations are recommended when vascular anomalies affecting the internal organs of the cavity are suspected. This is usually done when investigating the cause of occult bleeding. CVMs of the face and neck often require early pharyngo-laryngo-tracheoscopy because possible mucosal involvement can cause bleeding, infection, or airway complications. Conventional imaging methods may not be accurate enough to detect such damage. Endoscopic detection and destruction of the lesion can be achieved in the same session. Malformations located in the pelvic cavity and lower extremities often require proctosigmoidoscopy. Urethrocystoscopy and/or vaginoscopy (colposcopy) for early detection before bleeding. This is especially true for patients with CTS, which is associated with many gastrointestinal and genitourinary disorders. Arthroscopy is indicated for the evaluation of knee injuries, as small injuries are often not detected by conventional imaging methods. Accurate assessment is required for subsequent laser coagulation. Patients with BRBNS require regular esophagogastroduodenoscopy and full colonoscopy due to the high risk of gastrointestinal mucosal damage that can lead to severe bleeding. Perioral/intraoral HOI may require early pharyngo-larynx-tracheoscopic evaluation to identify an associated subglottic or tracheal hemangioma early before clinical symptoms appear. The vulvar lesion often needs a vaginoscopy/colposcopy and urethroscopy, while perianal lesion needs a proctoscopy.

### *Genetic Testing and Family Screening*

Germline and somatic mutations have been identified in a number of vascular anomalies, Sporadic syndromes may occur due to mosaic distribution of somatic mutations.

### *Specific Diseases:*

1. Hemangiomas

### *MRI is Indicated in Conditions like:*

1. Lesions consistent with PHACE syndrome (posterior fossa malformations, hemangiomas, arterial anomalies, coarctation of the aorta, cardiac defects, and eye anomalies) should undergo imaging to evaluate the carotids and cerebral vasculature.
2. The presence of multiple cutaneous lesions warrants screening with ultrasonography (US) or MRI to assess concomitant visceral lesions.
3. Lumbosacral lesions require imaging of the spinal cord (US or MRI) to rule out synchronous cord lesion.
4. Venous malformations.
5. Magnetic resonance angiography (MRA) or computed tomography (CT) angiography (CTA) may be required to delineate the full extent of complex venous malformations.

Such information may be useful for assisting in treatment and operative management.

6. Capillary malformations Imaging of the spinal cord should be done in the presence of capillary malformations; developmental defects of the central neural axis are common with these lesions.
7. Lymphatic malformations.
8. Large malformations of the lymph nodes can be diagnosed in utero. Such malformations are classified according to their radiographic and histological characteristics. Therefore, multimodal imaging is often used for appropriate delineation. MRI and Doppler US provide insight into extent and flow characteristics.
9. Arteriovenous malformations.

### *Treatment*

#### *Indications*

Treatment is based on the type of lesion, the associated symptoms, and the desires of the patient and family.

1. Most hemangiomas are small tumors that eventually subsides. However, treatment is necessary if the tumor is large, grows rapidly, aggravates a serious ulcer, is in a threatening location or may otherwise cause complications. Most of them are currently treated with propranolol as the first line of treatment.
2. Capillary malformations in patients who develop fibronodular hypertrophy or who have extensive facial involvement needs excision.
3. In venous malformations indications include appearance, impairment of function, and severe pain.
4. Lymphatic malformations treated for preventing infection and bleeding, cosmesis deformity, and improving the function of affected areas.
5. Quiescent arteriovenous malformations (AVMs) may be managed expectantly; however, pain, bleeding, ulceration, and extensive enlargement are all indications for treatment. Symptomatic lesions that are not amenable to surgical treatment may be considered for palliative embolization therapy.

## MEDICAL THERAPY

### *1. Hemangiomas*

Since 2008[17] and the consensus protocol report in 2013[18], propranolol has been used as first-line therapy for hemangiomas. Caution was exercised in infants with hemodynamic compromise, avoidance of hypoglycemia, and consideration that therapy outweighs the risks in patients with hemangioma associated with PHACE (posterior fossa, hemangioma, arterial, cardiac, eye) syndrome. The initial dose of propranolol is 0.5-1 mg/kg/day; if tolerated (heart rate and blood pressure measured after 1–3 doses) may be increased up to 2 mg/kg/day.[19]

Treatment of hemangiomas with corticosteroids and interferon alfanot preferred because of the common adverse effects (cushingoid syndrome for the former, neuropathy for the latter). Lauromacrogol has been used in sclerotherapy for refractory hemangioma.[20]

### *2. Venous Malformations*

Primary treatment for venous malformations consists of elastic compression devices to reduce swelling and pain. Daily aspirin may also be administered to prevent thrombotic complications. The main basis of treatment was sclerotherapy, mainly with ethanol.[21] Lauromacrogol was also used as a sclerosing agent in vascular malformations.[20] Sclerotherapy must be performed under general anesthesia, guided by ultrasound (US), fluoroscopy or both. With repeated treatment, the success rate of improving function and reducing symptoms can be up to 84%.[22]

### *3. Capillary Malformations*

Cosmetic camouflage and laser photocoagulation are the current first-line treatments.[23] Flashlamp pulsed dye lasers are most often used, though the results of such therapy are mixed. Multiple treatments are usually required, and nearly half of all lesions will darken within 5 years of treatment.[24]

Macrocytic lesions can be effectively treated with sclerotherapy. Both bleomycin and OK-432 (attenuated group A Streptococcus pyogenes) were effective as intralesional sclerosing agents.[25,26]

Burrows *et al*. Serio (2008) evaluated the use of doxycycline as an effective sclerosing agent in lymph node malformations.[27] Carbon dioxide, YAG laser therapy can also be used to treat mucosal lesions, although deformities treated with this therapy often recur and require repeated treatments.

### *4. Sclerotherapy*

In recent decades, sclerotherapy has become increasingly popular as a first-line treatment for lymphatic malformations due to its safety, efficacy, and cost-effectiveness compared to surgery; this development emphasizes the importance of interventional radiology in the multidisciplinary management of patients with these deformities.[28] The effectiveness and safety of sclerotherapy directly depends on the vascular architecture, the sclerosing agent used and the residence time of the sclerosant in the deformities.[29] The most commonly used sclerosing agents can be classified according to their mechanism of action[29] as follows:

- Direct induction of endothelial injury and thrombosis of the malformation - Ethanol, ethanolamine, and pingyangmycin
- Induction of a nonspecific inflammatory reaction within the malformation (less potent) - Sodium tetradecyl sulfate and bleomycin.

### *5. Newer Therapies*

More accurate diagnosis, improved treatment guidelines and various new forms of treatment are possible refinements in the classification and terminology of vascular disorders.[30] A number of molecular inhibitors targeting physiological pathways are being developed.[30] A newer agent that has been most studied to date is sirolimus [rapamycin[31]; mammalian target of rapamycin (mTOR) inhibitor], which is used to treat venous, lymph node, and complex malignancies. Another emerging agent is alpelizib (a PIK3CA inhibitor),[30] which is used in PIK3CA-mutated vascular malformations.

## SURGICAL THERAPY

### *1. Hemangiomas*

Surgical resection may be appropriate for lesions that are refractory to medical management. Resection may be performed at any of the three stages of the life cycle. As a general rule, resection should be deferred until the involuted phase (late childhood), when the lesion has matured and the anesthetic risk to the child is decreased. However, specific indications have been suggested for resection at all phases, as summarized by Marler and Mulliken:

- ***Infancy (proliferative phase)*** - Indications for resection include obstruction (visual or subglottic), bleeding, ulceration, deformity (eg, periorbital distortion), involvement of the scalp (to prevent alopecia of the effected region), and anticipation of a scar caused naturally involution
- ***Early Childhood (Involuting phase)*** - Large protuberant lesions are resected in this phase.
- ***Late childhood (Involuted phase)*** - Indications for resection in this phase include damaged skin, abnormal contour, and distortion of skin or surrounding structures

### *2. Venous Malformations*

Sclerotherapy is the primary treatment for venous malformations; but surgical excision may be offered for selected lesions. Small localized lesions are the best candidates for surgical intervention. Sclerotherapy should be used to shrink lesions prior to surgical excision.

### *3. Capillary Malformations*

Small fibrovascular lesions can easily be excised in most locations. More extensive excision and grafting of select capillary malformations may also be performed. Facial lesions with concomitant disfigurement may require excision with full or split-thickness grafts accompanied by contour resection and correction of maxillofacial distortion.[32,33]

### *4. Lymphatic Malformations*

Surgical resection is the mainstay of treatment for lymphatic malformations. In general, resection should be deferred until late infancy or early childhood to minimize anesthetic risk and to allow easier resection. Often, lymphatic malformations encompass vital structures, precluding complete excision. The approach to resection varies with lesion location. Generally, total excision of the lesion is attempted, with careful identification and preservation of involved major nerves (eg, preservation of the brachial plexus when an axillary lesion is excised). Resections tend to be involved,

and wound complications (eg, infection, drainage, swelling, and seroma formation) are common.

### 5. *Arteriovenous Malformations*

AVM treatment consists of a combination of embolization, sclerotherapy and surgical resection. Angiography is necessary for intervention, because it determines the extent of the damage and clearly identifies the feeding and drainage vessels. Surgical ligation of the feeding vessels should not be performed, as this will only cause new blood vessels to accumulate at the site of the lesion. Such a connection not only solves the problem, but even worse prevents endovascular access, thus avoiding therapeutic interventions.[23]

Embolization can be done with coils, particles or glue using arteries. Sclerotherapy can be given to the nidus vessels of the lesion simultaneously with occlusion of the feeding and draining vessels. Various sclerosing agents have been described [eg, absolute ethanol and N-butyl cyanoacrylate (NBCA)].[34] Combinations of embolization and sclerotherapy can be used to treat lesions that may not be operable; however, these procedures lead to only a temporary improvement, as new blood vessels are easily introduced into the lesion. All AVMs are high-flow lesions. Thus, it is necessary to ensure that the sclerosant does not escape from the lesion through the drainage vessels. This can lead to one of the most dire complications, non-target organ embolization, which can range from asymptomatic and clinically insignificant to problematic (eg, large skin wounds) or devastating (eg, blindness, organ failure or even fatal massive lung-resorption). If the lesion is considered suitable for surgery, preoperative embolization is usually recommended to facilitate resection. Certain lesions (eg, minor limb deformities) can be removed without preoperative embolization. Surgical procedures are designed to facilitate complete surgery and minimize blood loss and repeat surgery. Staging procedures are not usually performed, although for extensive, complex lesions, staged endovascular therapy (sclerosis, embolization, or plasticization) followed by single or staged surgical resection is an option.

The operation must involve the nidus of the lesion and any associated skin or deeper tissues. Wide excision is often necessary and the extent of resection is based on delineation of the lesion using preoperative imaging, bleeding patterns at the resection margin (ie, consistent with normal skin vasculature or extensive bleeding suggestive of deformity), and frozen sections resection margin. For major surgeries, grafting or tissue transfer may be necessary to ensure adequate wound closure. Fluorescence guided surgery with indocyanine green and infrared lamps has been recommended with varying results. Deep intracranial and complex intracranial AVMs present a unique therapeutic challenge, as surgical excision is often impossible. Embolization is the standard treatment for such lesions. Radiotherapy has also been successfully used to treat such intracranial lesions.[35]

### 6. *Combined Malformations*

Correct treatment of combined and complex deformities requires a high level of expertise in their treatment. Different parts of the deformity can be managed with separate therapeutic approaches. For a given patient, the multidisciplinary team may choose to embolize the arterial component of the lesion; sclerotherapy or surgical resection of a venous or lymphatic component; laser treatment of the skin component of the capillary; and staged surgical resection, mTOR inhibitors, or both for limb hyperplasia. Treatment must be individually adapted for each patient.

### *Laser*

The target of VM laser treatment is the pathologically dilated vascular endothelium. It is not light that the endothelial cells specifically absorb and emit heat. Satisfactory treatments cannot be expected if the treatment is not carried out by choosing the type of laser and changing the irradiation method based on the principles and limitations of phototherapy. For small-sized forms of the mucosa, tongue, lips and scalp, where scarring after treatment does not cause serious problems, there are several reports of resolution of lesions with the Nd:YAG laser.

### *Embolization*

The goal of AVM embolization is to destroy the nidus, and embolization in or near the nidus is necessary as much as possible. If feeding artery ligation/coil embolization is performed on the proximal/central side, the nidus does not disappear and the formation of multiple side channels is encouraged.

Resection is recommended within 3 days (72 h) after embolization. If the interval is prolonged, the risk of massive intraoperative bleeding may increase due to recanalization of the embolized

vessel and development of side channels. In addition, the lesion has been reported to worsen post-embolization injury.

### *Sclerotherapy*

Sclerotherapy for VM is effective in relieving symptoms and reducing the size of the lesion and is recommended. A range of complications have been reported, ranging from mild complications such as transient neuropathy and local inflammation to severe complications such as myopathy, skin necrosis, and deep vein thrombosis/pulmonary embolism. Sclerotherapy is generally considered effective in VM, but the problem is that there is little evidence and the procedure is not standardized. In addition, serious complications, which are rare but life-threatening, have been reported, and caution must be exercised when determining the dose of the sclerosing agent.

### *Conclusion*

ISSVA update on classification of vascular anomalies broadly divide it to vascular tumors and vascular malformations. The diagnostic evaluation methods are ultrasonography, CT and MRI with angiography along with newer radionucleotide scintigraphy. Management is multimodality with medical as well as surgical methods. Newer molecular targeted drugs are available. Sclerotherapy, cryotherapy and laser also developed for the management based on specific properties. In conclusion, the management of vascular malformations necessitates a holistic approach that combines clinical expertise, state-of-the-art imaging, and innovative treatment modalities. As our understanding of these complex anomalies continues to evolve, the goal remains the same: to provide optimal care that minimizes symptoms, improves patients' quality of life, and maximizes functional outcomes while minimizing the risk of complications.

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