# GUIDELINES

# Diagnosis and treatment of venous malformations Consensus Document of the International Union of Phlebology (IUP)-2009

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The International Union of Phlebology (IUP), the largest international organization devoted to the investigation and management of venous disorders, established an expert panel to formulate guidelines for physicians and health care professionals around the world on the evaluation and treatment of venous malformations (VMs).

The aim of this document is to provide recommendations for the diagnosis and treatment of VMs based on the best currently available scientific evidence. When scientific evidence was lacking or weak, a consensus of opinions among expert members of the panel was reached to support the recommendations.

The guidelines in this document are broad ranged and incorporate proven concepts and new discoveries. In the last decade, progress in both diagnostic techniques and minimally invasive technology has been significant in this difficult and challenging field. Imaging studies, radionuclide scans, ultrasound, computed tomography (CT) and magnetic resonance imaging (MRI) technologies have largely been perfected. The endovenous therapy revolution has transformed the way clinicians treat patients with venous disorders which includes VMs.

It is the sincere hope of the panel and the IUP, that these guidelines will serve its purpose: general guidelines based on scientific evidence to assist clinicians and patients in the diagnosis and treatment of VMs. The panel recognizes that some guidelines may be impractical in certain parts of the world with limited access to advanced technology or special expertise. To this end, the panel has incorporated the most important advances in this field to formulate the most up-to-date and sound guidelines based on the best available scientific evidence.

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## Definition of venous malformations

VMs are developmental anomalies (birth defects) of the venous system. They are the result of arrested development of the venous system during the

TABLE I.—*The modified Hamburg Classification of congenital vascular malformations primary classification.**

— Arterial malformations
— Venous malformations
— Arteriovenous malformations
— Lymphatic malformations
— Capillary malformation
— Combinedvascular malformations

Anatomical/embryological subclassification**
— Extratruncular forms
  diffuse, infiltrating
  limited, localized
— Truncular forms
  Obstruction or narrowing
  — aplasia; hypoplasia; hyperplasia
  Obstruction due to atresia or membranous occlusion
  Stenosis due to coarctation, spur, or membrane
  Dilatation
  — localized (aneurysm)
  Diffuse (ectasia)

* Based on the predominant vascular structure in the malformation; ** Based on anatomy and developmental arrest at the different stages of embryonal life: extratruncular form from earlier stages; truncular form from late stage.

various stages of embryogenesis. Together with arterial, capillary and lymphatic malformations they are part of a large group of congenital vascular malformations (CVMs) which are developmental anomalies of the peripheral vascular system [1-19] (Table I).

VMs should be differentiated from hemangiomas. Hemangiomas are vascular tumors that have a distinctly different etiology, genetics, presentation, prognosis and treatment (Mulliken and Glowacki). Hemangioma is a "self-limited" vascular tumor while CVMs are "self-perpetuating" embryologic tissue remnants. Precise understanding of this critical fact is required for successful CVM management and treatment.[20-34]

CVM remains a difficult diagnostic and therapeutic challenge among many vascular disorders due to the wide range of the clinical presentations, unpredictable clinical course, erratic response to the treatment with high recurrence/persistence rates, high morbidity following unspecific conventional treatment, and confusing terminology.[1-14]

CVM is, therefore, considered a unique vascular disorder that carries a stigma of totally unpredictable behavior. "Recurrence and persistence" is the trademark of all the CVMs. High recurrence rates are generally due to the embryological characteristics of the CVMs, which arise from embryonic tissue remnants derived from an earlier stage of embryogenesis. These lesions are now classified as "extratruncular" lesions by the Hamburg Classification.[7, 13, 35-43]

VM is the most common form among various CVMs. Most VMs exist alone as an independent lesion.

## Classification of venous malformations

Previous classification systems were established based purely on clinical findings. This was before the modern technology was available for accurate diagnosis. These classification systems failed to provide proper information concerning the etiology/embryology, anatomy, and pathophysiology involved in this vascular abnormality.

Numerous classifications of VMs have been proposed, many based on the appearance of the anomaly, its anatomy, pathology, or based on the velocity of blood flow in the lesion. Many VMs are still named after the clinician who first described the lesion. Lack of an accepted, universal classification system resulted in redundant terminology. For example, terminology such as "cavernous hemangioma", "cavernous angioma", "phlebangioma", "lymphangioma", "Port wine stain", etc. only added to the confusion.

Therefore, a new classification system has been accepted that fulfils the above criteria and provides information regarding lesion etiology, embryology, anatomy and pathophysiology of VMs.[15, 44-55]

*The Expert Panel of this document unanimously recommends the use of the Hamburg classification of CVMs, and within this the classification of VMs (Table I)*

The Hamburg classification [15, 44-47] was originally drafted based on a CVM Workshop held in Hamburg, Germany, organized by a group of specialists (Mulliken, Young, Belov and others) in 1988. The classification was further upheld and later modified by the then newly founded the International Society for the Study for Vascular Anomalies(ISSVA) in 1992, in Denver, CO, USA.

The original classification distinguished only four major-clinically and hemodynamically significant- groups of CVMs, and named them after the segment of the vascular tree that was the pre-

dominant lesion: arterial, venous, arteriovenous, and combined vascular malformations.

This classification was further modified with the addition of two more groups of malformations, the lymphatic and capillary malformations (Table I).

The Hamburg classification recognized that malformations are frequently mixed and venous or arteriovenous malformations may co-exist with lymphatic malformations or malformations of non-vascular tissue (bone, muscle, nerves, etc.). These malformations are classified as mixed CVMs. If the CVM lesion has elements of the lymphatic system, the term hemolymphatic malformation (HLM) is used.[56-69]

While the Panel discourages the use of eponyms in general, some mixed congenital malformations with well known eponyms have had a long history and tradition. A few of these eponyms are widely recognized today not only by vascular experts and societies, but by Foundations and the general public. One such malformation is known as Klippel-Trenaunay Syndrome (KTS), a mixed malformation with vascular, bony and soft tissue developmental anomalies.

The vascular malformations in KTS are usually mixed and include the venous, capillary and the lymphatic system. The term Parkes-Weber Syndrome is another widely accepted eponym used to describe high flow arteriovenous malformations that present frequently with other vascular malformations.[70-72]

In the Hamburg classification each CVM group is further subdivided into extratruncular and truncular forms, based on the embryological stage when developmental arrest has occurred.[15, 44-47]

### Extratruncular lesions

These are embryonic tissue remnants derived from an early stage of vascular tissue development (the reticular stage). Developmental arrest occurs before the main vascular trunks are formed (pretruncal embryonic lesions). These lesions maintain their unique embryonic characteristics of the mesenchymal cells and the ability to proliferate when stimulated by trauma, menarche, pregnancy, surgery, or other hormones. These lesions carry a high risk of recurrence compared with the much more frequently encountered truncular lesions.

Extratruncular lesions are further subdivided into diffuse, infiltrating and localized, limited lesions. Diffuse, infiltrating extratruncular lesions may cause symptoms due to compression of the surrounding structures (muscles, nerves). They may produce significant hemodynamic impact on the involved vascular system that is dependent on lesion size and location. Growth is usually slow and proportionate to the person's growth throughout the rest of the person's life. Furthermore, there is no spontaneous regression (cf. hemangioma).

### Truncular lesions

These lesions are the result of the developmental arrest that occurs during the "later" stages of vascular trunk formation during the fetal development. This arrest occurs long after the embryonic (reticular) stage of vascular development is over. These lesions are also known as "post-truncal fetal lesions".

Truncular lesions, therefore, do not have the embryonic characteristics of the mesenchymal cells (angioblasts) as observed in the extratruncular lesions. These lesions no longer possess the critical evolutional ability to proliferate. The risk of recurrence after treatment is minimal to none. These lesions have hemodynamic consequences due to congenital valvular incompetence, obstruction (atresia, hypoplasia) or dilatation/aneurysm formation with associated risk of thromboembolism.

Truncular lesions are subdivided into obstruction, aplasia or hypoplasia,[73-79] and dilation or aneurysms [79-91](Table I).

Immature/incomplete/abnormal development of the main axial veins result in aplasia, hypoplasia, or hyperplasia of the vessel (*e.g.*, agenesis/rudimentary femoral vein) or as a defective vessel: obstruction (*e.g.*, vein web, spur, annulus, or septum) or dilatation (*e.g.*, popliteal or iliac vein ectasia/aneurysm). These lesions also manifest as persistent, large, embryonic veins such as the marginal vein or the sciatic vein when a fetal (truncal) vessel fails to undergo normal involution.[92-95]

Truncular lesions of obstructive nature (webs, hypoplasia) may have different hemodynamic impacts on their relevant vascular systems depending upon their location, extent/severity, and natural compensation through collaterals. Chronic venous insufficiency develops in the territory drained by the truncular vein. Stenosing truncu-

TABLE II.—*ISSVA* classification of vascular anomalies, 1996, Rome, Italy.*

| Tumours | Vascular malformation simple | Combined |
|---|---|---|
| Hemagioma | Capillary malformation (CM) | CVM |
| Other tumors | Lymphatic malformation(LM) | CLVM |
| | Venous malformation (VM) | LVM |
| | Arteriovenous malformation (AVM) | CAVM |
| | | CLAVM |

*ISSVA: International Society for the Study for Vascular Anomalies.

lar lesions produce venous obstruction leading to a reduction in venous drainage. Membranous obstruction of the inferior vena cava in primary Budd-Chiari Syndrome is an example of a primary obstructive VM affecting a major vein.

Truncular VM lesions may also occur in veins with the same embryologic origin or draining the same territory (*e.g.*, stenosing lesions of the extracranial jugular veins, superior vena cava, and azygos vein system along the main outflow pathways of the cerebro-spinal venous system as suspected cause of multiple sclerosis).[96-99]

Avalvulia, or absence of valves is another form of hypoplasia that produces venous reflux. Together with atresia of the venous trunks and venous aneurysms, they are relatively common. The incidence of aneurysm has been reported to be 4% in nearly 490 cases of congenital anomalies of the venous system.[73]

Proper differentiation and recognition of the difference in embryological characteristics between "extratruncular" and "truncular" VM lesions is, therefore, critical. Proper classification as extratruncular or truncular is required for all the CVMs in order to ensure appropriate treatment and minimize the risk of recurrence.

This new (Hamburg) classification provided the impetus for the development of a contemporary concept of CVMs. With the Hamburg classification, precise diagnosis of various CVMs became feasible based on modern technology. Furthermore, a new concept of the 'multidisciplinary team approach' emerged aimed at the prevention and control of 'recurrence/persistence' with minimal possible complications and morbidity.

However, the current classification is far from the perfect and further modification will be necessary as our knowledge of the etiology, anatomy, embryology, histo-patho-physiology, hemodynamics, and possibly genetics of the CVMs continues to grow.

Further modification of the Hamburg Classification was proposed by ISSVA in 1996 to accommodate various pre-exisiting classifications (Table II). This revised classification included the vascular tumor (hemangioma) together with vascular malformations as a group of vascular anomalies. This distinction had limited value due to the complexity of the classification.

Venous malformations are further subclassified based on its anatomical location:

— intra-dermal, forming a superficial telangiectatic lesion;

— within the subcutaneous fat;

— intra-muscular, Intra-articular or deep within other organs.

VMs are also subclassified based on the clinical manifestations:

— localized : face, trunk, limbs, brain, spinal cord, lungs, etc.;

— generalized: Blue Rubber Bleb Syndrome, glomovenous malformations, genuine diffuse phlebectasisas (Bockenheimer)

## Diagnostic evaluation of venous malformations

### *Clinical evaluation*

Proper clinical evaluation of patients with VMs is essential. A thorough history, including a detailed birth and family history must be taken. The physical examination should include careful assessment (inspection, palpation, auscultation) of both the arterial and venous systems including a detailed pulse exam, making note of any edema, skin changes, varicosities, pigmentation, or ulcerations. An enlarged or longer extremity, digital

TABLE III.—*Duplex ultrasound assessment of vascular anomalies (VA).*

A dedicated vascular laboratory with expertise in the diagnosis of vascular anomalies should be performing these studies. Sonographers should be trained specifically in this field and should appreciate the complexity and the range of conditions they may encounter. Ultrasound assessment should be correlated with MRI findings. In case of deep intra-muscular lesions, the MRI may need to be done first to aid in locating the lesion on ultrasound. Assessment of the feeding and draining vessels is best done by venography.

An ultrasound study of VA should provide the following information:

*A. B-Mode*

- Gross ultrasonic morphology of the lesion and whether it is primarily composed of soft tissue (tumor) or vascular channels with little soft tissue (vascular malformation).
- The lesion measurements in length and cross-sectional diameter.[99]
- Location with respect to known landmarks.
- Location and depth of the lesion in the tissue (sub-cutaneous, intra-muscular, inter-muscular, peri-articular, intra-articular, etc.)
- Compressibilty of vascular channels and presence/absence of thrombus within the channels.
- Evidence of previous treatments (hyperechoic walls/segments), sclerothrombus, surgical scarring should be identified and commented on.
- Presence of other vessels in the vicinity and their contribution to the lesion. Normal anatomy should be identified and excluded. In case of arterial vessels, comparison with the contralateral side should be performed to make sure the vessel is part of the normal anatomy.
- In case of macrocystic lymphatic malformations, the size and number of cysts observed.

*B. Flow characteristics*

- Spectral, color and power Doppler examinations should confirm the flow characteristics.
- Flow characteristics (low flow vs. high flow) should be determined; assessment of flow direction under different postural and respiratory conditions should be included in the evaluation.[97, 98, 120-122]
- In case of VM involving the lower limbs, a separate venous incompetence study needs to be done to map the incompetent pathways. This is especially relevant when investigating complex malformations such as KTS.

*C. Other observations:*

Comments should be made regarding:

- Whether the lesion is unilateral or bilateral.
- If the underlying tissue shows hypertrophy, or atrophy.

anomalies and asymmetric growths of any part of the body must be recorded.

The appropriate combination of non-invasive to minimally- invasive tests should follow in order to confirm or exclude the clinical impression.[100-111]

### *Non-invasive tests*

Duplex scanning is the first test of choice for non-invasive evaluation of patients with VMs [112-119] (Table III):

— B-mode to differentiate tumors *vs.* malformations

— Doppler mode to assess flow characteristics

The panel recommends Duplex scanning as the first diagnostic test for all patients with VMs, involving the limbs, to assess the deep and superficial veins, to identify any aberrant vein, obstruction, dilation or valvular incompetence and define the feeding or draining veins of the VM. This test is safe, non-invasive, cost-effective and reliable (Grade of recommendation: 1-strong, level of evidence A-high quality).[56]

Duplex scanning is also useful in the assessment of the extracranial cerebral venous outflow [120-123] in addition to evaluation of aneurysms and stenoses of the jugular veins at cervical level.[83-91]

Other non-invasive studies, such as plethysmography, segmental pressure measurement, and pulse volume recordings should be used selectively and clinical correlations with abnormal findings (*e.g.*, outflow obstruction) need to be established.

Standard plain X-ray is still useful to identify abnormal findings in the soft tissue (*e.g.*, phlebolith) and other malformation-related abnormalities along the skeletal system.

### MINIMALLY INVASIVE TESTS

*Computed tomography with intravenous contrast.*—CT venography [124-128]is recommended for evaluation of obstructed veins and other truncu-

lar anomalies of large veins in the chest, abdomen or pelvis. Computed tomography accurately identifies the underlying pathology, confirms venous obstruction or extrinsic compression, delineates anatomic variations and extent of venous thrombosis (Grade of recommendation: 1-strong, level of evidence: B-moderate quality).[56]

*Magnetic resonance MRI and MR angiography* [129-138]

MRI and MR venography is recommended for evaluation of VMs. The test is reliable, it confirms the extent and type of the VM, delineates feeding and draining vessels, distinguishes between different soft tissues (muscle, fat) and the vascular structures. The imaging modality is highly accurate in the diagnosis of deep vein thrombosis. MRI and MRV is recommended before performing interventions on VMs, except some small localized VMs (Grade of recommendation: 1-strong, Level of evidence: A-high quality).[56]

The use of MR in infants and children, who would need anesthesia for the test should be selective and carefully planned.

*Whole body blood pool scintigraphy (WBBPS): transvenous angioscan utilizing radioisotope-tagged red blood cells.*—WBBPS[139-146] is an optional test to screen for multiple VM lesions scattered throughout the body. It allows qualitative and quantitative evaluation of the VM lesion especially during the course of multisession sclerotherapy as a cost-effective measure. It is an excellent tool for routine follow up and to assess the progress of treatment and the natural course of the VM lesion. It can exclude a combined VM-lymphatic malformation where the absence of an abnormal blood pool over the lymphatic lesion is the typical finding.

*Transarterial lung perfusion scintigraphy (TLPS): transarterial angioscan utilizing radioisotope-tagged microsphere albumin.*—TLPS [147-150]is not indicated for evaluation of the VM lesion. Its major function is to rule out the presence of a combined AV malformation (AVM) lesion. TLPS can detect micro-shunting of an AVM lesion which can be often be missed on conventional arteriography.

*Radionuclide lymphoscintigraphy* (LSG).—LSG [151-161] is essential to rule out lymphatic dysfunction especially due to the presence of a truncular lymphatic malformation known as primary lymphedema, which often occurs with the VM lesion.

— Microscopic fluorescent lymphagiography [162, 163]

— MR lymphangiography [164, 165]

— Ultrasound lymphangiography-investigational

— Endoscopy/colonoscopy for lesions involving the GI tract

*Invasive diagnostic tests*

a) Ascending, descending, and/or segmental venography/phlebography

b) Standard and/or selective arteriography

c) Percutaneous direct puncture angiography: arteriography, phlebography, varicography, lymphography

"Invasive" tests are seldom needed to establish the diagnosis of the VM and can be deferred until intervention is required. It is required for treatment planning either surgical or endovascular. However, invasive tests may be required for diagnosis when non- to minimally invasive tests (*e.g.*, CT and/or MRI) fail to confirm the diagnosis or to delinate important diagnostic details.

For example, an obstructive truncular VM lesion along the iliac vein often needs more precise anatomic information. Ascending phlebography combined with IVUS studies is essential for proper management. Descending phlebography is an indispensable tool to assess deep venous reflux along the pelvic veins and/or sciatic veins. These studies are required before treatment with embolotherapy. Direct puncture phlebography is also very useful to identify a large efferent vein of extratruncular lesions. These veins can be treated in advance to allow more effective therapy with reduced risk of recurrence, with subsequent embolotherapy or sclerotherapy.

*Blood tests*

Coagulation profile and D-dimer levels are also seldom indicated; extensive venous malformations and some vascular tumors are associated with a chronic form of disseminated intra-vascular coagulation. The following laboratory studies form an essential part of the patient's work-up:

— D-dimer- quantitative assay

— Fibrinogen

— Platelet count

— PT, APTT

— Thrombophlia screening for high risk malformations (*e. g.*, lesions involving the orbit).*

Whole blood count (WBC) - especially a measurement of hemoglobin in case of chronic blood loss *via* GI malformations.

*Histopathology*

A biopsy of the lesion should be performed to provide a histological diagnosis.[167-170] This is especially relevant when the differential diagnosis includes a non-involuting vascular tumour such as a non-involuting hemagioma (NICH). These lesions have high flow on Doppler and persist indefinitely and may be confused with an AVM.The evaluation of the majority of VMs can be achieved with the non- to minimally invasive tests alone.

*Immunohistochemistry*

The use of antisera anti-desmin/actin can delineate truncular defect of smooth muscle cell characteristic of primary venous aneurysm and other truncular VM.[83]

## Treatment of venous malformations

*Multidisciplinary team approach*

Surgical excision alone based on limited knowledge of the natural history and biology of the VM through earlier decades infrequently resulted in poor outcomes. These poor outcomes contributed to the confusion associated with the management of CVMs leading to mistaken prejudice.

But lately new endovascular therapies utilizing various forms of embolo/sclerotherapy were developed in order to improve the clinical outcome of extratruncular VM lesions. For truncular VM lesions, endovascular balloon dilatation and stenting techniques were also found to be beneficial in correcting the stenosing condition.

The new multidisciplinary team approach [171-183] aims for full integration of surgical, non-surgical and endovascular treatment options. This team concept is extended not only useful for diagnosis but is also essential for "combined" treatment using two or more different techniques. Surgical resection is typically combined with embolization. Furthermore, surgical resection may often require a vascular surgeon, a hand and/or plastic surgeon, or other specialists.

The multidisciplinary team often includes medical and allied health teams: Vascular Surgery, Pediatric Surgery, Plastic and Reconstructive Surgery, Orthopedic Surgery, Neurosurgery, Anesthesiology, Pathology, Physical Medicine and Rehabilitation, Oral-Maxillofacial Surgery, Head and Neck Surgery, Cardiovascular Medicine, Psychiatry, Dermatology, Interventional Radiology, Diagnostic Radiology, Nuclear Medicine, General Medicine, Neurology, Hematology, Genetics, General pediatrics, Occupational therapy, and many other health care practitioners.

The multidisciplinary team approach is also mandatory for proper selection/combination of the treatment modalities. All the decision related to the management should be based on the consensus among this multidisciplinary team approach as well as life-time follow up on the natural course and treatment outcomes.

## General principle

Not every VM lesion is amenable to treatment.[106-108, 184-186] Furthermore, not every VM lesion should be treated. Its mere presence often makes the practitioner feel obligated to treat. The only lesion assessed by the multidisciplinary team with justified indications should be considered for treatment. Although extratruncular VM lesions are more serious than truncular lesions with much poorer long-term outcome, an overzealous approach sometimes does more harm than good.[13]

"Not to intervene" is sometimes a wiser choice than to casually intervene without a full understanding of the biology and natural history of the VM lesion. Sometimes observation is the best approach. Another approach is to find an experienced center where the patient can be treated effectively in early childhood and not having to wait until after reaching adolescence.[187]

A "controlled" aggressive approach is favored where every effort is made to minimize collateral damage during treatment. In limb and life threatening situations, sacrificing limb over life may be necessary.

*In presence of thrombophilia and depending on the risk of the specific procedure, adequate anticoagulation should be provided.

The decision to initiate treatment should be based on the accepted indications.[188-197]

## General measures

*Explain the diagnosis*

An accurate diagnosis should be documented and communicated with the patient, parents or guardians of the pediatric patient and the referring doctor. Care should be taken to explain the difference between a tumor, such as a hemangioma, and a vascular malformation. This forms an integral part of educating the public and other medical specialties. Care should be taken to avoid confusing and redundant terminology such as "Port wine stain".

*Treat associated or secondary complications*

Examples include associated anemia from bleeding. Manage any associated pain and/or superficial recurrent thrombophlebitis.

*Graduated compression stockings and garments*

This is especially important for lesions involving the lower limbs. Compression garments are also useful for treatment of upper extremity VM lesions. Compression therapy can help with symptoms, which include edema and help prevent other complications such as superficial thrombophlebitis.

*Support and education*

Refer the patient to Support groups and recommend websites or print material to further educate the patient. Provide a referral for counseling or psychiatric assessment if required.

*Refer the patient to other specialists*

In case of leg length discrepancy, it is essential that children are referred early on to pediatric orthopedic surgeons, if vascular surgery alone or a combined treatment did not succeed in compensation of the length discrepancy.[198, 199-203] VM lesions involving the central nervous system require assessment by neurosurgeons and interventional neuroradiologists. Coagulation issues require consultation with a hematologist. Other specialists should be consulted as required. Allied health practitioners such as physiotherapists should also be involved and should form an integral part of the multi-disciplinary team approach.

*Family members screening and genetic counseling*

In cases of inherited malformations such as glomovenous malformations or blue rubber bleb syndrome, family member screening and genetic counseling may be indicated.

## Treatment

*General indications*

The panel strongly recommends that before treatment of any VMs the embryologic subtype of the VMs (extratruncular *vs.* truncular) be identified. The risks of thromboembolism, bleeding, injury to the surrounding structures (nerves, skin, bone, etc.) and likelihood of functional improvement and improved quality of life after potential treatment should be fully assessed. The presence of any other associated vascular malformations (arteriovenous shunting, lymphatic malformations) should also be determined. Careful assessment of the extent and severity of the VM lesion and identification of draining deep vein system is mandatory. This is especially true for truncular VM lesions of the lower extremity.[74, 192-194]

Indications for intervention may include the following conditions or complications of VMs:

— bleeding;

— signs and symptoms of chronic venous insufficiency (painful varicosity, edema, skin changes, ulcers, recurrent superficial thrombophlebitis);[195-196]

— lesions located at a life threatening region involving or close to vital structures (*e.g.*, proximity to the airway), or located in an area threatening vital functions (*e.g.*, sight, eating, hearing, or breathing);

— disabling pain:[197]

— functional impairment (*e.g.*, genital region);

— cosmetically severe deformity;

— lesions located at regions with high risk of complications (*e.g.*, hemarthrosis, thromboembolism);

— lesions combined producing the vascular-bone syndrome (length discrepancy of the lower

extremities, affecting the bone itself) [187, 198-203] or the destructive angiodysplastic arthritis(Hauert disease);[245]

— lesions obstructing the outflow and drainage of vital organ (*i.e.*, liver, brain);[76, 77, 204-207]

— persistent lymph leak due to a combined lymphatic malformation lesion with/without infection;[208]

— recurrent sepsis, local and/or general, due to a combined lymphatic malformation lesion.[209-213]

*Treatment strategy*

When the benefit of treatment outweighs the risk of complications and morbidity, less risky treatment options (*e.g.*, foam/liquid sclerotherapy) should be first line therapy. "No treatment is the best option if feasible". In contrast to the treatment of AVMs, all VM lesions can be treated using a less aggressive approach.[34, 185, 214-216]

The traditional conservative approach to the young pediatric patient with a VM is still valid, especially for the common VMs without bony involvement. 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.

Development of the vascular-bone syndrome with resultant long bone length discrepancy or the destructive angiodysplastic arthritis with resultant immobility, and the presence of a VM lesion at a life or limb threatening anatomic location, are situations where an earlier treatment approach is preferred over a more conservative one.[12, 13, 173, 217]

Orthopedic manipulation of the non-affected limb to correct a leg length discrepancy should be discouraged.[12, 13]

In the presence of a life or limb-threatening, condition (*e.g.*, hemorrhage) treatment should be started expeditiously despite the risk of the associated morbidity.

*Treatment modalities*

OBSERVATION AND CONSERVATIVE MANAGEMENT

Many asymptomatic and small lesions are best managed with observation or conservative, compression treatment. The panel suggests a conservative approach to most asymptomatic lesions and recommends any treatment other than of very small, localized VMs be performed only by vascular specialists, most frequently after multidisciplinary consultations (Grade of recommendation: 1-strong, level of evidence: B-moderate quality).[56]

Conservative approach also includes proper skin care, local treatment of bleeding or ulcerative lesions and drug therapy of complications like superficial thrombophlebitis.

DRUG THERAPY

There is no specific drug to improve/control the VM lesions in contrast to the infantile/neonatal hemangioma. Anticoagulation is often required to treat thrombotic complications and resultant morbidity associated with VM lesions.

ENDOVASCULAR THERAPY

Endovascular therapy (*e.g.*, ethanol sclerotherapy) is now a universally accepted independent therapy of VMs in the poor surgical candidate with extensive lesions extending beyond the deep fascia with involvement of muscle, tendon and bone as seen in diffuse infiltrating extratruncular lesions.

ETHANOL SCLEROTHERAPY

Ethanol is a potent irritant sclerosant causing trans-mural destruction of the vessel.[218-224] Ethanol is the only proven sclerosant available that can deliver near-complete control of the nidus of any extratruncular CVM when utilized appropriately and is associated with excellent long term outcomes.

Ethanol sclerotherapy requires special training and experience in order to minimize the risk of complication and subsequent morbidity. This agent should be used only discriminately in the treatment of VM and LM.

Ethanol sclerotherapy has become the gold standard sclerotherapy agent by which all other agents are compared.

Ethanol is also considered the best agent for sclerotherapy of the "diffusely infiltrating" extratruncular VM lesion. In experienced hands, the risk of complications is low and recurrence is rare. Unfortunately, complications can be severe if ethanol is injected close to large nerves or into the skin.

Ethanol sclerotherapy has a high rate of com-

plications and morbidity if the VM is located in the lip, tongue, gum/oral mucosa or in the hand at fingers, or in the foot at the toe, or palm, sole with or without transdermal extension. VM lesions with transdermal extension or in close proximity to the skin or mucosa are known to carry a high risk of skin or mucosa necrosis.

Because the majority of VM lesions are seldom life or limb threatening, the 'indiscriminate' use of the ethanol to treat all VM lesions has been called into question.

### Sclerotherapy with other liquid sclerosants

Before the era of the ethanol, various liquid sclerotherapy agents have been used in the treatment of VM lesions over the past several decades often resulting in high recurrence rates and poor long term results.[225-231]

Ethibloc and polidocanol are the two most popular agents that have been widely used in Europe for several decades. These agents are not yet available in the US. Ethibloc is an emulsion made of viscous ethanol and corn protein but its mechanism of action is mechanical occlusion followed by intravascular fibrosis. It carries a high risk of non-target vascular occlusion due to its viscosity.

In the US sodium tetradecyl sulfate (STS) and ethanolamine oleate have been used with limited success in the treatment of VM lesions. Because of the high morbidity associated with ethanol, STS remains the major scelosant in the treatment of VM lesions.

### Ultrasound-guided sclerotherapy with foam sclerosants

Because of the high morbidity associated with the use of ethanol in the treatment of CVMs, interest in the development and utilization of detergent based sclerosants (*e.g.*, STS, polidocanol) for the treatment of VMs, resulted in a new treatment approach based on the foam sclerotherapy.[232-237]

Ultrasound guided foam sclerotherapy using polidocanol or STS can deliver satisfactory results with minimal morbidity, when used to treat high risk 'localized' lesions. A preliminary assessment of this treatment modality demonstrated lesion recurrence (35% with two year follow-up), which was amenable to repeat foam sclerotherapy.

Foam sclerotherapy is a treatment option in a selected group of 'diffuse infiltrating' VM lesions which would otherwise be treated with ethanol. The associated risk of collateral damage seen with ethanol sclerotherapy (*e.g.*, nerve injury, muscle contraction), can largely be avoided with foam sclerotherapy. Foam sclerotherapy can deliver good relief of symptoms and clinical improvement with minimal risk of complications, in this extended group of infiltrating VM lesions.

Higher recurrence remains the major disadvantage of foam sclerotherapy compared with ethanol sclerotherapy. But the foam sclerotherapy produces excellent short to mid-term control of small VM lesions. Long term assessment of foam sclerotherapy outcomes is required in order determine if these results apply to all types of VM lesions.

Foam sclerotherapy of vascular malformations requires proper training and experience. Given the interaction of detergent sclerosants with plasma proteins, coagulation, antithrombotic, fibrinolytic and other physiological systems, the unknown fluid mechanics and undefined rheology of foams in large low flow embryonic vascular spaces, and given the possibility of drainage into the central venous system, and the potential for systemic complications and cerebrovascular events, we caution against the use of large volumes of foam sclerosants in treating extensive infiltrating VM lesions. The recommended maximum safe volume of foam, based on local and international standards, should be not be exceeded.

### Fluoroscopic and ultrasound guided sclerotherapy (FUGS)

Combining sonographic and fluoroscopic guidance to deliver sclerosants in the treatment of venous malformations has a particular relevance to the treatment of venous malformations.[238, 239]

Ultrasound guidance is used to identify and localize the target vessel(s), contrast medium is then injected allowing visualization of the target lesion and the draining veins on fluoroscopy. STS or polidocanol (POL) foam is then introduced slowly into the lesion which appears radio-lucent on fluoroscopy displacing most of the radio-opaque contrast agent. The injection is stopped when the draining veins to take up the foam sclerosant. Compression is applied and maintained for seven days post-operatively.

Fluoroscopy allows a more comprehensive visualization of the target lesion and draining veins which is otherwise not possible with ultrasound imaging alone.

FUGS is particularly useful in treatment of intramuscular venous malformations.

### Embolotherapy with coils, glue, and/or particles embolization

Currently available embolization agents are not ideal for VM lesions since these lesions are generally low flow and high volume lesions with large diameter vascular channels.[148, 225] Micro-particles and coils are usually not large enough to occlude such lesions effectively and are often washed out.

Embolization agents are unable to produce complete destruction of the vessel endothelium. Incomplete endothelial cell destruction carries a significant risk of lesion recurrence. Furthermore, these agents only produce mechanical compression of the lesion and cessation of flow that results in thrombosis.

The role of endovascular therapy is therefore, relatively limited with the exception of N-butylcyanoacrylate embolotherapy (NBCA). NBCA is ideal as an adjunctive agent used to fill up the VM lesion preoperatively to facilitate surgical excision and reduce the risk of bleeding. NBCA improves the safety and effectiveness of surgical excision and reduces the risk of bleeding.

### Endovenous thermal ablations (laser, radiofrequency, cryoablation)

Endoluminal thermal ablation may have a complementary role in small, limited VM lesions.[240, 241]

These new ablative techniques have demonstrated efficacy in the treatment of venous incompetence and are currently being assessed in treatment of venous malformations. The findings so far are encouraging but more detailed studies are needed to further assess the efficacy of these new modalities.

### Angioplasty and stent

Angioplasty and stenting has been shown to be efficacious in the treatment of obstructive iliac vein and vena caval lesions. This endovascular approach is also useful for treating stenosing truncular VM lesions: webs, septum, and stenosis of the iliac vein, inferior vena cava, jugular vein, and azygous vein, and to relieve chronic venous hypertension.[75, 76, 173, 204, 206]

### Open surgical therapy

Open surgical therapy combined with the endovascular therapy (embolo/sclerotherapy), is the most effective means to control VM lesions.[53, 74, 186, 188, 193, 194, 214, 242-244] Active incorporation of the embolo/sclerotherapy pre- and/or post-operatively allows substantial expansion of the traditional role of surgical excisional. This is especially true for the infiltrating extratruncular VM lesion.[172, 174, 177, 197, 217]

Surgical excision is the treatment of choice for truncular VM lesions which fail to respond to endovascular therapy.[78]

Among the various surgical procedures available for the treatment of VM lesions, vascular procedures to correct hemodynamic derangements (venous hypertension) should have a priority. Examples include reconstructive surgery (*e.g.*, venous bypass) and ablative surgery (*e.g.*, removal of marginal vein; excision/removal of vascular defects).

Non-vascular (non-hemodynamic) operations aiming to correct the secondary consequences of VM should be deferred until appropriate primary vascular procedures are performed. Examples of non-vascular operations include orthopedic surgery (*e.g.*, Achilles tendon lengthening) and plastic and reconstructive surgery to correct cosmetic deformities.[198, 199, 202, 203]

A "combined" surgical approach is also preferred in situations where other surgical specialists are needed such as neurosurgeon, urologist, plastic surgeon, etc.

### *Follow-up*

The patient or patient's parents/guardian should be informed of the post operative requirements. All possible symptoms which may be of concern post treatment should be described and the patient or parents/guardian is instructed to contact the treating practitioner urgently if they arise. Practitioner follow-up of the parent's or guardian's concerns should be appropriate addressed in a timely manner. Contact details of the treating phlebologist should be provided.

The follow-up processes should include:

— An assessment of the treatment should be made by examining the patient on the following occasions:

— 1 week- An ultrasound DVT scan follow up if indicated in high risk patients;

— 6-12 weeks after the completion of the course of treatments, a duplex study should be organized to assess the effectiveness of the procedure and look for persistence/early recurrence;

— 6-12 months after the completion of the course of treatments, a repeat duplex ultrasound +/- a follow-up MRI should be arranged to assess the effectiveness of the treatment and look for persistence/late recurrence.

According to the indicators at each assessment the findings should be recorded and discussed with the patient or parents/guardian and include:

— success of treatment including resolution of symptoms;

— degree of sclerosis and any recannalisation;

— any complications;

— patient satisfaction.

Reviewing processes to include:

— where clinically indicated, further appropriate treatment is offered, or referral made;

— treatment and assessment records are complete and include:

location of the lesion treated;

treatment parameters used including sclerosant form, concentration and volume and laser fluence, power and energy density;

diameter of the lesion(s) treated;

type and size of compression garment applied and recommended time of application;

Post-treatment assessment of resolution of symptoms;

after post-treatment assessments, what further treatment is indicated/offered (*e.g.*, ultrasound guided sclerotherapy or direct vision sclerotherapy) (if any), any referral made, and whether the treatment plan has been completed;

— any adverse effects or interventions and their resolutions

*Complications*

Superficial thrombophlebitis (STP), deep vein thrombosis (DVT) and pulmonary embolism (PE)

— Involving vital or critical structures

eyes and extension into brain

perineum, genitals

intra-articular

— Thrombosis and calcification. Disseminated intravascular coagulopathy (DIC)

— Chronic venous hypertension, Lipodermatosclerosis and ulceration

— Limb hypertrophy, scoliosis and other orthopaedic abnormalities.

## Conclusions

Multidisciplinary approach with full integration of open surgical and endovascular therapy has become the mainstay of treatment in the contemporary management of venous malformations.

A team approach using new treatment strategies can improve the long-term treatment outcomes and reduce the morbidity and recurrence/persistence rates compared with conventional approaches.

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