GUIDELINES

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

B. B. LEE [1], J. BERGAN [2], P. GLOVICZKI [3], J. LAREDO [1], D. A. LOOSE [4], R. MATTASSI [5], K. PARSI [6], J. L. VILLAVICENCIO [7], P. ZAMBONI [8] *

[1]*Division of Vascular Surgery, Department of Surgery, Center for Vein, Lymphatics, and Vascular Malformation, Georgetown University School of Medicine, Washington, DC, USA*
[2]*Department of Surgery, UCSD School of Medicine, University of California, San Diego, CA, USA*
[3]*Division of Vascular and Endovascular Surgery, Department of Surgery, Mayo Clinic College of Medicine, Director, Gonda Vascular Center, Mayo Clinic, Rochester, MN, USA*
[4]*Department for Vascular Surgery, European Centre for the Diagnosis and Treatment of Vascular Malformations, Die Facharztklinik Hamburg, Hamburg, Germany*
[5]*Department of Vascular Surgery, Stefan Belov Center for Vascular Malformation, G. Salvini Hospital, Garbagnate Milanese, Milan, Italy*
[6]*University of New South Wales, Australia and Australian College of Phlebology, Sydney, Australia*
[7]*Department of Surgery, Uniformed Services University School of Medicine, and Venous and Lymphatic Teaching Clinic, Walter Reed Army Medical Center, Bethesda, MD, USA*
[8]*Vascular Diseases Center, University of Ferrara, Ferrara, Italy*

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.

[Int Angiol 2009;28:434-51]

*Editorial Committee

Chairman: B. B. Lee, MD, PhD, FACS

Members: J. J. Bergan, MD, FACS, FRCS, P. Gloviczki, MD, FACS, J. Laredo, MD, PhD, D. A. Loose, MD, R. Mattassi, MD, K. Parsi, MD, FACD, FACP, J. Lionel Villavicencio, MD, P. Zamboni, MD

Received on November 1, 2009; accepted for publication on November 31, 2009

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.

References

1. Malan E, Puglionisi A. Congenital angiodysplasias of the extremities, note II: arterial, arterial and venous, and hemolymphatic dysplasias. J Cardiovasc Surg (Torino) 1965;6:255-345.
2. DeTakats G. Vascular anomalies of the extremities. Surg Gynecol Obstet 1932;55:227-37.
3. Tasnadi G. Epidemiology and etiology of congenital vascular malformations. Semin Vasc Surg 1993;6:200-3.
4. Laor T, Burrows PE. Congenital anomalies and vascular birthmarks of the lower extremity. Magn Reson Imaging Clin N Am 1998;6:497-519.
5. Enjolras O, Mulliken JB. Vascular malformations. In Harper J, Oranje A, Prose N. Textbook of pediatric dermatology. Oxford: Blackwell Sciences Ltd; 2000. p. 975-7.
6. Vollmar J, Vogt K. Angiodysplasie und Skeletsystem. Der Chirurg 1976;47:205-13.
7. Belov ST, Loose DA, Weber J, editors. Vascular malformations. Periodica Angiologica XVI. Reinbek b. Hamburg: Einhorn-Presse; 1989. p. 138-200.
8. Villavicencio JL. Congenital vascular malformations: historical background. Phlebology 2007;22:247-48.
9. Mattassi R. Historical background. In: Mattassi R, Loose DA, Vaghi M, editors. Hemangiomas and vascular malformations. An atlas of diagnosis and treatment. Milan: Springer Verlag; 2009. p. 9-13.
10. Lee BB. Advanced management of congenital vascular malformation (CVM). International Angiology 2002;21: 209-13.
11. Lee BB. Current concept of venous malformation (VM). Phlebolymphology 2003;43:197-203.
12. Lee BB, Kim HH, Mattassi R, Yakes W, Loose D, Tasnadi G. A new approach to the congenital vascular malformation with a new concept: how the pioneer Prof. Stefan Belov enlightened us through the Seoul consensus. Int J Angiol 2003;12:248-51.
13. Lee BB, Mattassi R, Loose D, Yakes W, Tasnadi G, Kim

HH. Consensus on Controversial Issues in Contemporary Diagnosis and Management of Congenital Vascular Malformation – Seoul Communication. Int J Angiol 2004;13:182-92.
14. Lee BB. Changing concept on vascular malformation: no longer enigma. Annals of Vascular Diseases 2008;1:11-9.
15. Lee BB, Laredo J, Lee TS, Huh S, Neville R. Terminology and classification of congenital vascular malformations. Phlebology 2007;22:249-52.
16. Loose DA. Vascular malformations. Surgery 1997;15:39-43.
17. Lee BB. Congenital venous malformation: changing concept on the current diagnosis and management. Asian J Surgery 1999;22:152-4.
18. Malan E. Hemodynamic phenomena in congenital arteriovenous fistulae. In: Malan E, editor. Vascular malformations. Milan: Carlo Erba; 1974. p. 30-7.
19. Malan E. History and nosography. In: Malan E, editor. Vascular malformations (Angiodysplasias). Milan: Carlo Erba; 1974. p. 15-9.
20. Boon LM, Enjolras O, Mulliken JB. Congenital hemangioma: evidence of accelerated involution. J Pediatr 1996;128:329-35.
21. Enjolras O, Riché MC, Merland JJ, Escandej P. Management of alarming hemangiomas in infancy: a review of 25 cases. Pediatrics 1990;85:491-8.
22. Enjolras O, Wassef M, Mazoyer E, Frieden IJ, Rieu PN, Drouet L *et al.* Infants with Kasabach-Merritt syndrome do not have "true" hemangiomas. J Pediatr 1997;130:631-40.
23. Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. Plast Reconstr Surg 1982;69:412-22.
24. Mulliken JB. Classification of vascular birthmarks. In: Mulliken JB, Young AE, editors. Vascular birthmarks, hemangiomas and malformation. Philadelphia, PA: WB Saunders; 1988. p. Finn MC, Glowacki J, Mulliken JB. Congenital vascular lesions: clinical application of a new classification. J Pediatr Surg 1983;18:894-900.
25. Jackson IT, Carreno R, Potparic Z, Hussain K. Hemangiomas, vascular malformations, and lymphovenous malformations: classification and methods of treatment. Plast Reconstr Surg 1993;91:1216-30.
26. Stal S, Hamilton S, Spira M. Hemangiomas, lymphangiomas, and vascular malformations of the head and neck. Otolaryngol Clin North Am 1986;19:769-96.
27. Waner M, Suen JY. The natural history of hemangiomas. In: Waner M, Suen JY, editors. Hemangiomas and vascular malformations of the head and neck. New York, NY: Wiley-Liss; 1999. p. 13-46.
28. Byard RW, Burrows PE, Izakawa T, Silver MM. Diffuse infantile hemangiomatosis: Clinicopathologic features and management problems in five fatal cases. Eur J Pediatr 1991;150:224-7.
29. Mulliken JB, Zetter BR, Folkman J. In vivo characteristics of endothelium from hemangiomas and vascular malformations. Surgery 1982;92:348-53.
30. Mulliken JB. Cutaneous vascular anomalies. Semin Vasc Surg 1993;6:204-18.
31. Mulliken JB. Classification of vascular birthmarks. In: Grainger RG, Allison DJ, editors. Vascular birthmarks, haemangiomas, and malformations. Philadelphia, PA: WB Saunders; 1988. p. 24-37.
32. Mulliken JB. Treatment of hemangiomas. In: Mulliken JB, Young AE, editors. Vascular birthmarks, hemangiomas and malformations. Philadelphia, PA: WB Saunders; 1988. p. 88-90.
33. Loose DA. Vascular malformations and hemangiomas: clinical features and their basis. In: Chang JB, editor. Textbook of angiology. New York, NY: Springer; 2000. p. 1248-57.
34. Szilagyi DE, Elliott JP, DeRusso FJ, Smith RF. Peripheral congenital arteriovenous fistulas. Surgery 1965;57:61-81.
35. Szilagyi DE, Smith RF, Elliott JP, Hageman JH. Congenital arteriovenous anomalies of the limbs. Arch Surg 1976;111:423-9.
36. Lee BB. Critical issues on the management of congenital vascular malformation. Annals Vasc Surg 2004;18:380-92..
37. Woolard HH. The development of the principal arterial stems in the forelimb of the pig. Contrib Embryol 1922;14:139-54.
38. Lewis FT. Development of the veins in the limbs of rabbit embryos. Am J Anat 1906;5:113-20.
39. Leu HJ. Pathoanatomy of congenital vascular malformations. In: Belov S, Loose DA, Weber J, editors. Vascular malformations. Reinbek, Germany: Einhorn-Presse Verlag; 1989. p. 37-46.
40. Bastide G, Lefebvre D. Anatomy and organogenesis and vascular malformations. In: Belov St, Loose DA, Weber J, editors. Vascular malformations. Reinbek: Einhorn-Presse Verlag GmbH; 1989. p. 20-2.
41. Dickinson P. Venous stasis and bone growth. Exp Med Surg 1953;11:49-53.
42. Acsady G, Solti F, Frank J, Turbok E. The development of extremital arteriovenous shunts after deep venous thrombosis. In: Maurer PC, Becker HM, Heidrich H *et al.*, editors. What is new in angiology. Proceedings of the 14th World Congress of the International Union of Angiology. Munich, Germany: W. Zuckschwerdt Verlag; 1986. p. 457-9.
43. Belov S. Classification, terminology, and nosology of congenital vascular defects. In: Belov S, Loose DA, Weber J, editors. Vascular malformations. Reinbek, Germany: Einhorn-Presse; 1989. p. 25-30.
44. Belov S. Anatomopathological classification of congenital vascular defects. Semin Vasc Surg 1993;6:219-24.
45. Rutherford RB. Classification of peripheral congenital vascular malformations. In: Ernst C, Stanley J, editors. Current therapy in vascular surgery. 3rd ed. St. Louis, MO: Mosby; 1995. p. 834-8.
46. Belov S. Classification of congenital vascular defects. Int Angiol 1990;9:141-6.
47. Van Der Stricht J. Classification of vascular malformations. In: Belov St, Loose DA, Weber J, editors. Vascular malformations. Reinbek: Einhorn-Presse Verlag GmbH; 1989. p. 23.
48. Puig S, Casati B, Staudenherz A, Paya K. Vascular low-flow malformations in children: current concepts for classification, diagnosis and therapy. Eur J Radiol 2005;53:35-45
49. Gloviczki P, Duncan AA, Kalra M, Oderich GS, Ricotta JJ, Bower TC *et al.* Vascular malformations: an update. Perspect Vasc Surg Endovasc Ther 2009;21:133-48.
50. Mattassi R, Loose DA, Vaghi M. Classification of vascular malformations. In: Mattassi R, Loose DA, Vaghi M, editors. Hemangiomas and vascular malformations. An atlas of diagnosis and treatment. Milan: Springer Verlag; 2009.
51. Lee BB. New approach to old problem of venous disease-congenital vascular malformation. In: Angelides NS, editor. Advances in phlebology. Limassol: Hadjigeogiou Printing & Co.; 1998. p. 59-64.
52. Belov ST, Loose DA, Mattassi R, Spatenka J, Tasnadi G, Wag Z. Therapeutical strategy, surgical tactics and operative techniques in congenital vascular defects (Multicentre Study). In: Strano A, Novo S, editors. Advances

in vascular pathology. Vol. 2. Amsterdam, New York, Oxford: Excerpta Medica; 1989. p. 1355-60.
53. Orvidas LJ, Kasperbauer JL. Pediatric lymphangiomas of the head and neck. Ann Otol Rhinol Laryngol 2000;109:411-21.
54. Berwald C, Salazard B, Bardot J, Casanova D, Magalon G. Port wine stains or capillary malformations: surgical treatment. Ann Chir Plast Esthet 2006;51:369-72.
55. Summary of Guideline of American Venous Forum. Handbook of venous disorders. 3rd edition. In: Gloviczki P., editor. London, UK: A Hodder Arnold; 2008. p. 702-7.
56. Klippel M, Trenaunay I. Du naevus variqueux et ostéohypertrophique. Arch Gén Méd 1900:3:641-72.
57. Servelle M. Klippel and Trénaunay's Syndrome. Ann Surg 1985:201:365-73.
58. Conaway CW, Villavicencio JL, Gomez ER, Coffey JA, Salander JM, Rich NM. The surgical management of the Klippel-Trenaunay syndrome. Abstract. The American Venous Forum; 1989.
59. Villavicencio JL, Conaway CW, Pikoulis E, Gannon MX. Congenital vascular malformations of venous predominance. The Klippel Trenaunay Syndrome. In: Raju S, Villavicencio JL, editors. Surgical management of venous disease. Media, PA: Williams and Wilkins; 1997. p. 445-67.
60. Villavicencio JL. Congenital vascular malformations - predominantly venous? The Syndrome of Klippel-Trenaunay. Scope on Phlebology and Lymphology 2000;71:116-25.
61. Jacob AG, Driscoll DJ, Shaughnessy WJ, Stanson AW, Clay RP, Gloviczki P. Klippel-Trenaunay syndrome: spectrum and management. Mayo Clin Proc 1998;73:28-36.
62. 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.
63. Noel AA, Gloviczki P, Cherry KJ Jr, Rooke TW, Stanson AW, Driscoll DJ. Surgical treatment of venous malformations in Klippel-Trenaunay syndrome. J Vasc Surg 2000;32:840-7.
64. Gloviczki P, Hollier LH, Telander RL, Kaufman B, Bianco AJ, Stickler GB. Surgical implications of Klippel-Trenaunay syndrome. Ann Surg 1983;197:353-62.
65. Lee A, Driscoll D, Gloviczki P, Clay R, Shaughnessy W, Stans A. Evaluation and management of pain in patients with Klippel-Trenaunay syndrome: a review. Pediatrics 2005;115:744-9.
66. Servelle M. Klippel and Trénaunay's Syndrome. 768 operated cases. Ann Surg 1985;201:365-73.
67. Dragieva G, Stahel HU, Meyer M, Kempf W, Haffner A, Burg G, Hafner J. Proteus syndrome. Vasa 2003;32:159-63.
68. Servelle M, Babillot J. Deep vein malformations in the Klippel-Trenaunay syndrome Phlebologie 1980;33:31-6.
69. Gloviczki P, Driscoll DJ. Klippel-Trenaunay syndrome: current management. Phlebology 2007; 22:291-8.
70. Weber FP. Angioma formation in connection with hypertrophy of limbs and hemihypertrophy. Brit J Derm Syph 1907:19:231-5.
71. Ziyeh S, Spreer J, Rossler 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.
72. Eifert S, Villavicencio JL, Kao TC, Taute BM, Rich NM. Prevalence of deep venous anomalies in congenital vascular malformations of venous predominance. J Vasc Surg 2000;31:462-71.
73. Belov ST. Congenital agenesis of the deep veins of the lower extremity: surgical treatment. J Cardiovasc Surg 1972:13:594.
74. Raju S, Hollis K, Neglen P. Obstructive lesions of the inferior vena cava: clinical features and endovenous treatment. J Vasc Surg 2006;44:820.
75. Lee BB, Villavicencio L, Kim YW *et al.* Primary Budd-Chiari syndrome: outcome of endovascular management for suprahepatic venous obstruction. J Vasc Surg 2006;43:101-10.
76. Valla DC. Primary Budd-Chiari syndrome. J Hepatol 2009;50:195-203.
77. Zamboni P, Pisano L, Mari C, Galeotti R, Feo C, Liboni A. Membranous obstruction of the inferior vena cava and Budd-Chiari syndrome. Report of a case. J Cardiovasc Surg (Torino) 1996;37:583-7.
78. Nedelmann M, Kaps M, Mueller-Forell W. Venous obstruction and jugular valve insufficiency in idiopathic intracranial hypertension. J Neurol 2009 [Epub ahead of print].
79. Leriche H, Aubin ML, Aboulker J. Cavo-spinal phlebography in myelopathies. Stenoses of internal jugular and azygos veins, venous compressions and thromboses Acta Radiol Suppl 1976;347:415-7.
80. Gillespie DL, Villavicencio JL, Gallagher C, Chang A, Hamelink JK, Fiala LA *et al.* Presentation and management of venous aneurysms. J Vasc Surg 1997;26:845-52.
81. Villavicencio JL, Gillespie DL, Eifert S. Venous aneurysms: presentation and treatment. In: current therapy in vascular surgery. 4th edition. In: Ernst CB, Stanley JC, editors. Philadelphia, PA: Mosby Publ.; 1999.
82. Zamboni P, Cossu A, Carpanese L, Simonetti G, Massarelli G, Liboni A. The so-called venous aneurysms. Phlebology 1990;5:45-50.
83. Friedman SG, Krishnasastry KV, Doscher W, Deckoff SL. Primary venous aneurysms. Surgery 1990;108:92-5.
84. Andreev A, Petkov D, Kavrakov T, Penkov P. Jugular venous aneurysms: when and how to operate. Int Angiol 1998;17:272-5.
85. Kersting S, Rössel T, Hinterseher I, Gaebler R, Litz R, Bergert H *et al.* Isolated aneurysm of the internal jugular vein. Vasa 2008;37:371-3.
86. Bush S, Khan R, Stringer MD. Anterior jugular venous aneurysm. Eur J Pediatr Surg 1999;9:47-8.
87. Ilijevski NS, Radak S, Novakovic B, Miholjcic A, Radak D. Images in vascular medicine. Jugular vein aneurysm--ultrasonographic evaluation. Vasc Med 2006;11:51.
88. Fishman G, DeRowe A, Singhal V. Congenital internal and external jugular venous aneurysms in a child. Br J Plast Surg 2004;57:165-7.
89. Lee HY, Yoo SM, Song IS, Yu H, Lee JB. Sonographic diagnosis of a saccular aneurysm of the internal jugular vein. J Clin Ultrasound 2007;35:94-6.
90. Ilijevski NS, Radak S, Vucureviç G, Sagiç D, Otaseviç P, Tasiç N *et al.* Jugular vein aneurysm. Vascular 2008;16:291-4.
91. Vollmar JF, Voss E. Vena marginalis laterlis persistens – die vergessene Vene der Angiologen. Vasa 1979;8:192.
92. Mattassi R. Approach to marginal vein: current issue. Phlebology 2007;22:283-6.
93. Kim YW, Lee BB, Cho JH, Do YS, Kim DI, Kim ES. Haemodynamic and clinical assessment of lateral marginal vein excision in patients with a predominantly venous malformation of the lower extremity. Eur J Vasc Endovasc Surg 2007;33:122-7.
94. Weber J, Daffinger N. Congenital vascular malformations: the persistence of marginal and embryonal veins. Vasa 2006;35:67-77.
95. Tzuladze II. The selective phlebography of the large tributaries of the vena cava system in the diagnosis of venous circulatory disorders in the spinal complex. Zh Vopr Neirokhir Im N N Burdenko 1999;2:8-13.
96. San Millan Ruiz D, Gailloud P, Rufenacht DA, Delavel-

le J, Henry F *et al*. The craniocervical venous system in relation to cerebral venous drainage. AJNR Am J Neuroradiol 2002;23:1500-8.
97. Gisolf J, van Lieshout JJ, van Heusden K, Pott F, Stok WJ, Karemaker JM. Human cerebral venous outflow pathway depends on posture and central venous pressure. J Physiol 2004;560(Pt 1):317-27.
98. Zamboni P, Galeotti R, Menegatti E, Malagoni AM, Tacconi G, Dall'Ara S *et al*. Chronic cerebrospinal venous insufficiency in patients with multiple sclerosis. J Neurol Neurosurg Psychiatry 2009;80:392-9.
99. Lee BB, Laredo J, Deaton DH, Neville RF. Arteriovenous malformations: evaluation and treatment. Chapter 53. Handbook of venous disorders: Guidelines of the American Venous Forum. 3rd edition. Gloviczki P, editor. London: UK A Hodder Arnold; 2009. p. 583-93.
100. Lee BB. Vascular surgery; cases, questions and commentaries. van Urk G, Hobson II C, editors. Chapter 40. Congenital vascular malformation. London: Springer-Verlag Limited; 2003. p. 315-23.
101. Lee BB, Beaujean M. Nouvelles strategies dans la prise en charge des malformations vasculaires congenitales (MVC): un apercu de l'experience clinique coreenne. Angeiologie 2004;56:11-25.
102. Lee BB, Laredo J, Lee SJ, Huh SH, Joe JH, Neville R. Congenital vascular malformations: general diagnostic principles. Phlebology 2007;22:253-7.
103. Villavicencio JL. Investigation of congenital malformations. In: Nicolaides AN, editor. Investigation of chronic venous insufficiency. A Consensus Statement. Circulation 2000;102:e126-e163:27-8.
104. Villavicencio JL. Classification/critical diagnostic steps: How deep does it go? In: Venous/arterial malformations: a difficult and challenging problem with some hope. Miami, FL: Post Graduate Course, American Venous Forum Annual Meeting; 2006 Feb 22-25. p. 28-9.
105. Mattassi R, Colombo R, Boccalon M, Vaghi M, D'Angelo F, Tacconi A. Experiences in the surgical treatment of congenital vascular malformations; changes in diagnosis and surgical tactics in the view of new experiences. In: Belov ST, Loose DA, Weber J, editor. Vascular malformations. Reinbeck: Einhorn Presse Verlag; 1989. p. 202-5.
106. Mattassi R, Vaghi M. Vascular bone syndrome-angiosteodystrophy. Current Concept Phlebology 2007; 22:287-90.
107. Mattassi R. Diagnosis and treatment of venous malformations of the lower limbs. In: Wang ZG, Becker HM, Mishima Y, Chang J. Vascular surgery. Beijing, China: International Academic Publishers; 1993. p. 397-404.
108. Rutherford RB. New approaches to the diagnosis of congenital vascular malformations. In: Belov S, Loose DA, Weber J, editors. Vascular malformations. Vol. 16. Reinbek, Germany: Einhorn-Presse Verlag; 1989. p. 60-5.
109. Rutherford RB. Congenital vascular malformations: diagnostic evaluation. Semin Vasc Surg 1993;6:225-32.
110. Stillo F. Diagnostics and treatment of peripheral venous malformations: state of the art. Angéiologie 1998;50: 30-40.
111. Lee BB, Mattassi R, Choe YH, Vaghi M, Ahn JM, Kim DI *et al*. Critical role of Duplex ultrasonography for the advanced management of a venous malformation (VM). Phlebology 2005;20:28-37.
112. Dubois J, Patriquin HB, Garel L, Powell J, Filiatrault D, David M, Grignon A. Soft-tissue hemangiomas in infants and children: diagnosis using Doppler sonography. AJR Am J Roentgenol 1998;171:247-52.
113. 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.
114. Timmerman D, Wauters J, Van Calenbergh S, Van Schoubroeck D, Maleux G, Van Den Bosch T *et al*. Color Doppler imaging is a valuable tool for the diagnosis and management of uterine vascular malformations. Ultrasound Obstet Gynecol 2003;21:529-31.
115. Mahesh B, Thulkar S, Joseph G, Khazanchi RK, Srivastava A. Color Duplex ultrasound-guided sclerotherapy: a new approach to the management of patients with peripheral vascular malformations. Clin Imaging 2003;27:171-9.
116. Hubsch P, Trattnig S, Kainberger FM, Barton P, Karnel F. Imaging of peripheral arteriovenous malformations using color-coded Doppler sonography. Ultraschall Med 1991;12:87-90.
117. Paltiel HJ, Burrows PE, Kozakewich HP, Zurakowski D, Mulliken JB. Soft-tissue vascular anomalies: utility of US for diagnosis. Radiology 2000;214:747-54.
118. Offergeld C, Schellong SM, Daniel WG, Huttenbrink KB. Value of color-coded duplex ultrasound in interstitial laser therapy of hemangiomas and vascular malformations Laryngorhinootologie 1998;77:342-6.
119. Menegatti E, Zamboni P. Doppler haemodynamics of cerebral venous return. Curr Neurovasc Res 2008;5: 260-5.
120. Valdueza JM, von Munster T, Hoffman O, Schreiber S, Einhaupl KM. Postural dependency of the cerebral venous outflow. Lancet 2000;355:200-1.
121. Schreiber SJ, Lurtzing F, Gotze R, Doepp F, Klingebiel R, Valdueza JM. Extrajugular pathways of human cerebral venous blood drainage assessed by Duplex ultrasound. J Appl Physiol 2003;94:1802-5.
122. Vargel I, Çil BE, Kiratli P, D'Akinci, Erk Y. Hereditary intraosseous vascular malformation of the craniofacial region: imaging findings. Br J Radiol 2004;77:197-203
123. Hyodoh H, Hori M, Akiba H, Tamakawa M, Hyodoh K, Hareyama M. Peripheral vascular malformations: imaging, treatment approaches, and therapeutic issues. RadioGraphics 2005;25:S159-S71.
124. Rubin PAD, Bilyk JR, Dunya IM, Weber AL. Spiral CT of an orbital venous malformation. AJNR 1995;16: 1255-7.
125. Elsayes KM, Menias CO, Dillman JR, Platt JF, Willatt JM, Heiken JP. Vascular malformation and hemangiomatosis syndromes: spectrum of imaging manifestations. Am J Roentgenol 2008;190:1291-9.
126. Elsayes KM, Menias CO, Dillman JR, Platt JF, Willatt JM, Heiken JP. Vascular Malformation and Hemangiomatosis Syndromes: spectrum of imaging manifestations. AJR 2008;190:1291-9.
127. Napoli A, Fleischmann D, Chan FP, Catalano C, Hellinger JC, Passariello R *et al*. Computed tomography angiography: state-of-the-art imaging using multidetectorrow technology. J Comput Assist Tomogr 2004;28(Suppl 1):S32-45.
128. Lee BB, Choe YH, Ahn JM, Do YS, Kim DI, Huh SH *et al*. The new role of MRI (Magnetic Resonance Imaging) in the contemporary diagnosis of venous malformation: can it replace angiography? J Am Coll Surg 2004;198: 549-58.
129. Rak KM, Yakes WF, Ray RL, Dreusbach JN, Parker SH, Luethke JM *et al*. MR imaging of symptomatic peripheral vascular malformations. AJR Am J Roentgenol 1992;159:107-12.
130. Baker LL, Dillon WP, Hieshima GB, Dowd CF, Frieden IJ. Hemangiomas and vascular malformations of the head and neck: MR characterization. AJNR Am J Neuroradiol 1993;14:307-14.
131. Gelbert F, Riche MC, Reizine D, Guichard JP, Assouline

E, Hodes JE *et al*. MR imaging of head and neck vascular malformations. J Magn Reson Imaging 1991;1: 579-84.
132. Smith JK, Castillo M, Wilson JD. MR characteristics of low-flow facial vascular malformations in children and young adults. Clin Imaging 1995;19:109-17.
133. Herborn CU, Goyen M, Lauenstein TC, Debatin JF, Ruehm SG, Kroger K. Comprehensive time-resolved MRI of peripheral vascular malformations. AJR Am J Roentgenol 2003;181:729-35.
134. Dobson MJ, Hartley RW, Ashleigh R, Watson Y, Hawnaur JM. MR angiography and MR imaging of symptomatic vascular malformations. Clin Radiol 1997;52: 595-602.
135. Kern S, Niemeyer C, Darge K, Merz C, Laubenberger J, Uhl M. Differentiation of vascular birthmarks by MR imaging. An investigation of hemangiomas, venous and lymphatic malformations. Acta Radiol 2000;41:453-7.
136. Yonetsu K, Nakayama E, Miwa K, Tanaka T, Araki K, Kanda S *et al*. Magnetic resonance imaging of oral and maxillofacial angiomas. Oral Surg Oral Med Oral Pathol 1993;76:783-9.
137. Lo Casto A, Salerno S, Cannizzaro F, Caronia A, Bencivinni F, Barbiera F *et al*. MRI findings in lingual venous malformations. Dentomaxillofac Radiol 2003;32:333-6.
138. Lee BB, Kim BT, Choi JY, Cazaubon M. Prise en charge des malformations vasculaires congénitales (MVC) en 2003: rôle de la scintigraphy corps entier dans las surveillance évolutive. Angeiologie 2003;55:17-26.
139. Lee BB, Mattassi R, Kim BT, Kim DI, Ahn JM, Choi JY. Contemporary diagnosis and management of venous and AV shunting malformation by whole body blood pool scintigraphy (WBBPS). Int Angiol 2004;23:355-67.
140. Hollerman JJ, Bernstein MA, Froelich JW, Schkudor G. Detection of hemangiomas using whole-body imaging with technetium-99m labeled RBCs. Clin Nucl Med 1986;11:716-7.
141. Inoue Y, Wakita S, Ohtake T, Ohkubo T, Hayashi N, Nishikawa J *et al*. Use of whole-body imaging using Tc-99m RBC in patients with soft-tissue vascular lesions. Clin Nucl Med 1996;21:958-9.
142. Fukuda Y, Murata Y, Umehara I, Yamashita T, Ono C, Iwai T *et al*. Perfusion and blood pool scintigraphy for diagnosing soft-tissue arteriovenous malformations. Clin Nucl Med 1999;24:232-4.
143. Front D, Israel O, Groshar D, Weininger J. Technetium-99m-labeled red blood cell imaging. Semin Nucl Med 1984;14:226-50.
144. Barton DJ, Miller JH, Allwright SJ, Sloan GM. Distinguishing soft-tissue hemangiomas from vascular malformations using technetium-labeled red blood cell scintigraphy. Plast Reconstr Surg 1992;89:46-52; discussion 53-5.
145. Sloan GM, Bolton LL, Miller JH, Reinisch JF, Nichter LS. Radionuclide-labeled red blood cell imaging of vascular malformations in children. Ann Plast Surg 1988;21:236-41.
146. Lee BB, Mattassi R, Kim BT, Park JM. Advanced Management of Arteriovenous Shunting Malformation (AVM) with transarterial lung perfusion scintigraphy (TLPS) for follow-up assessment. Int Angiol 2005;24:173-84.
147. Lee BB. Mastery of vascular and endovascular surgery. In: Zelenock H, Messina L, Moneta editors). Chapter 76. Arteriovenous malformation. p 597-607. Philidelphia, PA: Lippincott, Williams and Wilkins; 2006.
148. Lee BB, Laredo J, Deaton D, Neville R. Endovascular treatment of some congenital diseases - hemangioma and vascular malformation. In: Heuser RR, Henry M, editors. Textbook of peripheral vascular interventions. Chapter 82. Section XI. 2nd edition. London: Informa Healthcare; 2008. p. 712-22.
149. Lee BB. Congenital vascular malformation. In: van Urk G, Hobson II C, editors. Vascular surgery. Chapter 41. 2nd edition. Cases, questions and commentaries. London: Springer-Verlag; 2006. p. 377-92.
150. Lee BB. Lymphatic malformation. Chapter 4. Lymphedema diagnosis and treatment. In: Tredbar, Morgan, Lee, Simonian, Blondeau, editors. London: Springer-Verlag; 2008. p. 31-42.
151. Lee BB, Kim YW, Seo JM, Hwang JH, Do YS, Kim DI *et al*. Current concepts in lymphatic malformation (LM). Vasc Endovasc Surg 2005;39:67-81.
152. Lee BB. Lymphedema-Angiodysplasia Syndrome: a prodigal form of lymphatic malformation (LM). Phlebolymphology 2005;47:324-32.
153. Lee BB. Lymphedema-diagnosis and treatment. In: Tredbar, Morgan, Lee, Simonian, Blondeau, editors. Lymphatic malformation. Chapter 4. Springer-Verlag London Limited 2008.
154. Baulieu F, Baulieu JL, Vaillant L, *et al*. Factorial analysis in radionuclide lymphography: assessment of the effects of sequential pneumatic compression. Lymphology 1989;22:178-85.
155. Choi JY, Hwang JH, Park JM, Lee KH, Kim SE, Kim DI *et al*. Risk assessment of dermatolymphangioadenitis by lymphoscintigraphy in patients with lower extremity lymphedema. Kor J Nucl Med 1999;33:143-51.
156. Choi JY, Lee KH, Kim SE, Kim BT, Hwang JH, Lee BB. Quantitative lymphoscintigraphy in post-mastectomy lymphedema: correlation with circumferential measurements (abstract). Kor J Nucl Med 1997;31:262.
157. McNeill GC, Williams WH, Witte MH, Witte CL. Whole body lymphangioscintigraphy. Lymphology 2002;35 (Suppl):219-22.
158. Williams WH, McNeill GC, Witte MH, Witte CL. Evaluation of peripheral lymphedema by longitudinal lymphangioscintigraphy. Lymphology 2002;35(Suppl):223-7.
159. Witte CL, Witte MH, Unger EC, Williams WH, Bernas MJ, McNeill GC *et al*. Advances in imaging of lymph flow disorders. RadioGraphics 2000;20:1697-719.
160. Lee BB, Bergan, JJ. New clinical and laboratory staging systems to improve management of chronic lymphedema. Lymphology 2005;38:122-9.
161. Allegra C, Sarcinella R, Bartolo M Jr. Morphologic and functional changes of the microlymphatic network in patients with advancing stages of primary lymphedema. Lymphology 2002;35:114-20.
162. Franzeck UK, Spiegel I, Fischer M, Bortzler C, Stahel HU, Bollinger A. Combined physical therapy for lymphedema evaluated by fluorescence microlymphography and lymph capillary pressure measurements. J Vasc Res 1997;34:306-11.
163. Harisinghani MG, Dixon WT, Saksena MA, Brachtel E, Blezek DJ, Dhawale PJ *et al*. MR Lymphangiography: Imaging Strategies to Optimize the Imaging of Lymph Nodes with Ferumoxtran-101 RadioGraphics 2004;24:867-78.
164. Lohrmann C, Felmerer G, Foeldi E, Bartholomä JP, Langer M. MR lymphangiography for the assessment of the lymphatic system in patients undergoing microsurgical reconstructions of lymphatic vessels. Microvasc Res 2008;42-5.
165. Lee BB. Regarding non-contrast three-dimensional magnetic resonance imaging vs lymphoscintigraphy in the evaluation of lymph circulation disorders: A comparative study. J Vasc Surg 2005;4:821-2.
166. Leu HJ. Pathomorphology of vascular malformations: analysis of 310 cases. Intern Angiol 1990;9:147-55.

167. Leu HJ. Zur Morphologie der arteriovenösen Anestomosen bei kongenitalen Angiodysplasien. Morphol Med 1982;2:99-107.
168. Stein JA, Heidary N, Pulitzer M, J Schaffer JV, North P. Noninvoluting congenital hemangioma. Dermatol Online J 2008;14:7.
169. Zamboni P, Menegatti E, Galeotti R, Malagoni AM, Tacconi G, Dall'Ara S *et al*. The value of cerebral Doppler venous haemodynamics in the assessment of multiple sclerosis. J Neurol Sci 2009 [Epub ahead of print].
170. Lee BB, Bergan JJ. Advanced management of congenital vascular malformations: a multidisciplinary approach. J Cardiovascular Surgery 2002;10:523-33.
171. Loose DA. Combined treatment of congenital vascular defects: indications and tactics. Semin Vasc Surg 1993;6:260-5.
172. Lee BB, Laredo J, Kim YW, Neville R. Congenital vascular malformations: general treatment principles. Phlebology 2007;22258-63.
173. Loose DA. Angeborene Gefäßmalformationen. In: Alexander K, editors. Gefäßkrankheiten. München: Urban und Schwarzenberg; 1994.
174. Loose DA. Malformaciones vasculares. Sistemática para el diagnósticó radiológico y la terapéutica. Forum FL 1997;2:101-8.
175. Lee BB. Critical role of multidisciplinary team approach in the new field of vascular surgery – endovascular surgery. J Kor Soc Vasc Surg 2003;19:121-3.
176. Loose DA. Combined treatment of vascular malformations: indications, methods and techniques. In: Chang JB, editor. Textbook of angiology. New York, NY: Springer; 2000. p. 1278-83.
177. Lee BB. Vascular malformations. J Am Coll Surg 2005;200:638-9.
178. Villavicencio JL, Scultetus A, Lee BB. Congenital vascular malformation: when and how to treat them. Semin Vasc Surg 2002;15:65-71.
179. Lee BB. Malformazioni vascolari ed emangiomi. Trattamento delle angiodisplasie di tipo venoso. Capitolo 19. p. 116-123. Milano: Springer-Verlag Italia; 2003.
180. Loose DA. Contemporary treatment of congenital vascular malformations. In: Dieter RS, Dieter Jr RA, Dieter RA III, editor. Peripheral arterial disease. New York, NY: McGrawHill; 1993. p. 1025-40.
181. Lee BB. New approaches to the treatment of congenital vascular malformations (CVMs) – Single center experiences – (Editorial Review). Eur J Vasc Endovasc Surg 2005;30:184-97.
182. Mattassi R. Multidisciplinary treatment. In: Mattassi R, Loose DA, Vaghi M, editors. Vascular malformations and hemangiomas. An atlas of diagnosis and treatment. Milano: Springer; 2009.
183. Mattassi R. Treatment of venous malformations. In: Mattassi R, Loose DA, Vaghi M, editors. Hemangiomas and vascular malformations. An atlas of diagnosis and treatment. Milan: Springer Verlag; 2009. p. 223-30.
184. Villavicencio JL. Primum non nocere: Is it always true? The use of absolute ethanol in the management of congenital vascular malformations. J Vasc Surg 2001;33: 1-3.
185. Mattassi R. Surgical treatment of congenital arteriovenous defects. Int Angiol 1990;9:196-202.
186. Soltesz L. Contributions of clinical and experimental studies of the hypertrophy of the extremities in congenital arteriovenous fistulae. Proceedings of the 7th Congress of the International Cardiovascular Society. J Cardiovas Surg (Special Supplement Issue) 1965;5.
187. Belov St, Loose DA. Surgical treatment of congenital vascular defects. Int Angiol 1990;9:175-82.
188. Lee BB. Klippel-Trenaunay syndrome and pregnancy. Intern Angiol 2003;22:328.
189. Lee BB, Laredo J. Letter to the Editor - Multidisciplinary approach in the management of a giant arteriovenous malformation in the right axillary region. J Vasc Surg 2008;48:775-6.
190. O'Donovan JC, Donaldson JS, Morello FP, Pensler JM, Vogelzang RL, Bauer B. Symptomatic hemangiomas and venous malformations in infants, children, and young adults: treatment with percutaneous injection of sodium tetradecyl sulfate. AJR Am J Roentgenol 1997;169: 723-9.
191. Belov ST. Late results of surgical treatment of congenital vascular defects. In: Maurer PC, Becker HM, Heidreich H, Hoffmann G, Kriessmann A, Müller-Wiefel H, Prätorius C, editors. What is new in angiology? Trends and controversies. München, Bern, Wien: Zuckerschwerdt; 1986. p. 249-50.
192. Belov ST. Operative-technical peculiarities in operations of congenital vascular defects. In: Balas P, editor. Progress in angiology. Torino: Minerva Medica; 1992. p. 379-82.
193. Belov ST. Vascular malformations and hemangiomas: surgical treatment. In: Chang JB, editor. Textbook of angiology. p. 1284-93.
194. Hulsmanns RFHJ. Congenital angiodysplastic Syndromes associated with primary or secondary varicosis and/or phlebectasias. Scope on Phlebology and Lymphology 1995;2:8.
195. Villavicencio JL. Treatment of varicose veins associated with congenital vascular malformations. In: Bergan JJ, Goldman MP, editors. Complex problems involving varicose veins. Part 4. Varicose veins and telangiectasias. St Louis, MO: Quality Medical Publ; 1993. P. 329-42.
196. Loose DA. Modern tactics and techniques in the treatment of angiodysplasias of the foot. Chir del piede 2001;25:1-17.
197. Belov ST. Haemodynamic pathogenesis of vascular bone syndromes in congenital vascular defects. Int Angiol 1990;9:155-61.
198. Belov ST. Correction of lower limbs length discrepancy in congenital vascular bone disease by vascular surgery performed during childhood. Semin Vasc Surg 1993;6:245-51.
199. Kim YW, Do YS, Lee SH, Lee BB. Risk factors for leg length discrepancy in patients with congenital vascular malformation. J Vasc Surg 2006;44:545-53.
200. Weber FP. Angioma formation in connection with hypertrophy of limbs and hemihypertrophy. Brit J Derm Syph 1993;19:231-5.
201. Mattassi R. Differential diagnosis in congenital vascular-bone syndromes. Semin Vasc Surg 1993;6:233-44.
202. Mattassi R, Vaghi M. Vascular Bone Syndrome – angioosteo-dystrophy: current concepts. Phlebology 2007;22:287-90.
203. Zamboni P, Galeotti R, Menegatti E, Malagoni AM, Mascoli F, Dall'Ara S *et al*. Rationale and preliminary results of endovascular treatment of multiple sclerosis, the liberation procedure. In: Greenhalgh RM. Vascular and endovascular controversies update. London: BIBA Publishing; 2009. p. 71-8.
204. Hirooka K, Hirooka M, Kisaka Y, Uehara T, Hiasa Y, Michitaka K *et al*. Doppler waveform pattern changes in a patient with primary Budd-Chiari syndrome before and after angioplasty. Intern Med 2008;47:91-5.
205. Lee BB, Laredo J, Deaton D, Neville R. Endovascular management of Budd-Chiari Syndrome – Suprahepatic Inferior Vena Cava Occlusive Disease. In: Heuser RR, Henry M, editors. Textbook of peripheral vascular interventions. Chapter 83. Section XII. 2nd edition. London: Informa Healthcare; 2008. p. 725-31.
206. Xu K, He FX, Zhang HG, Zhang XT, Han MJ, Wang CR *et al*. Budd-Chiari syndrome caused by obstruction of the hepatic inferior vena cava: immediate and 2-year

treatment results of transluminal angioplasty and metallic stent placement. Cardiovasc Intervent Radiol 1996;19:32-6.
207. Tasnadi G, Mattassi R, Fox U. Chyledema of a limb and chylothorax treated successfully with a Denver shunt. Lymphology 1998;31(Suppl):381-4.
208. Lee BB, Kim DI, Whang JH, Lee KW. Contemporary management of chronic lymphedema – personal experiences. Lymphology 2002;35(Suppl):450-5.
209. Park JH, Kim DI, Huh S, Lee SJ, Do YS, Lee BB. Absolute ethanol sclerotherapy on cystic lymphangioma in neck and shoulder region. J Korean Vascular Surgery Society 1998;14:300-3.
210. Kim KH, Kim HH, Lee SK, Seo JM, Chang WY, Lee BB. OK-432 intralesional therapy for lymphangioma in children. J Korean Association of Pediatric Surgeons 2001;7:142-6.
211. Lee BB, Laredo J, Seo JM, Neville R. Hemangiomas and vascular malformations. In: Mattassi, Loose, Vaghi, eidtors. Chapter 29. Treatment of lymphatic malformations. Milano: Springer-Verlag Italia; 2009. p. 231-50.
212. Hancock BJ, St-Vil D, Luks FI, Di Lorenzo M, Blanchard H. Complications of lymphangiomas in children. J Pediatr Surg 1992;27:220-4; discussion 224-6.
213. Loose DA. Surgical management of venous malformations. Phlebology 2007;22,6:276-82.
214. Blei F. Congenital lymphatic malformations. Annals of the New York Academy of Sciences 2009;1131:185-94.
215. Lim SW, Kim DI, Huh S, Lee SJ, Do YS, Lee JS *et al*. Clinical experiences with combination of selective embolization and surgery for treatment of congenital vascular malformation. J Korean Vascular Surgery Society 1998;14:304-8.
216. Loose DA, Weber J. Indications and tactics for a combined treatment of congenital vascular defects. In: Balas P, editor. Progress in angiology. Torino: Edizione Minerva Medica; 1991. p. 373-8.
217. Lee BB, Kim DI, Huh S, Kim HH, Choo IW, Byun HS *et al*. New experiences with absolute ethanol sclerotherapy in the management of a complex form of congenital venous malformation. J Vasc Surg 2001;33:764-72.
218. Lee BB, Do YS, Byun HS, Choo IW, Kim DI, Huh SH. Advanced management of venous malformation (VM) with ethanol sclerotherapy: mid-term results. J Vasc Surg 2003;37:533-8.
219. Jeon YH, Do YS, Shin SW, Liu WC, Cho JM, Lee MH *et al*. Ethanol embolization of arteriovenous malformations: results and complications of 33 cases. J Kor Radiol Soc 2003;49:263-70.
220. Shin BS, Do YS, Lee BB, Kim DI, Chung IS, Cho HS *et al*. Multistage ethanol sclerotherapy of soft-tissue aretriovenous malformations: effect on pulmonary arterial pressure. Radiology 2005;235:1072-7.
221. Do YS, Yakes W, Shin SW, Lee BB. Ethanol embolization of arteriovenous malformations: interim results. Radiology 2005;235:674-82.
222. Lee BB. Fast facts-vascular surgery highlights 2006-07. Chapter: Management of Arteriovenous Malformation. Abington, Oxford: Health Press Limited; 2007. p. 42-50.
223. Rimon U, Garniek A, Galili Y, Golan G, Bensaid P, Morag B. Ethanol sclerotherapy of peripheral venous malformations. Eur J Radiol 2004;52:283-7.
224. Rosenblatt M. Endovascular management of venous malformations. Phlebology 2007;22:264-75.
225. Mimura H, Kanazawa S, Yasui K *et al*. Percutaneous sclerotherapy for venous malformations using polidocanol under fluoroscopy. Acta Med Okayama 2003;57:227-34. Avram J [Allergic shock after injection of sodium tetradecyl sulfate]. Phlebologie 1966;19:157-8.
226. Siniluoto TM, Svendsen PA, Wikholm GM, Fogdestam I, Edstrom S. Percutaneous sclerotherapy of venous malformations of the head and neck using sodium tetradecyl sulphate (sotradecol). Scand J Plast Reconstr Surg Hand Surg 1997;31:145-50.
227. Riche MC, Hadjean E, Tran-Ba-Huy P, Merland JJ. The treatment of capillary-venous malformations using a new fibrosing agent. Plast Reconstr Surg 1983;71:607-14.
228. Dubois JM, Sebag GH, De Prost Y, Teillac D, Chretien B, Brunelle FO. Soft-tissue venous malformations in children: percutaneous sclerotherapy with Ethibloc. Radiology 1991;180:195-8.
229. Coleridge Smith P. Saphenous ablation: sclerosant or sclerofoam? Semin Vasc Surg 2005;18:19-24.
230. Gelbert F, Enjolras O, Deffrenne D, Aymard A, Mounayer C, Merland JJ. Percutaneous sclerotherapy for venous malformation of the lips: retrospective study of 23 patients. Neuroradiology 2000;42:692-6.
231. Lee BB, Bergan J. Transition from alcohol to foam sclerotherapy for localized venous malformation with high risk. Chapter 12. A textbook-foam sclerotherapy. In: Bergan J, Van Le Cheng, editors. London: The Royal Society of Medicine Press Ltd; 2008. p. 129-39.
232. Bergan J, Pascarella L, Mekenas L. Venous disorders: treatment with sclerosant foam. J Cardiovasc Surg (Torino) 2006;47:9-18.
233. Pascarella L, Bergan JJ, Yamada C, Mekenas L. Venous angiomata: treatment with sclerosant foam. Ann Vasc Surg 2005;19:457-64.
234. Bergan J, Cheng V. Foam sclerotherapy of venous malformation. Phlebology 2007;22:299-302.
235. Yamaki T, Nozaki M, Fujiwara O, Yoshida E. Duplex-guided foam sclerotherapy for the treatment of the symptomatic venous malformations of the face. Dermatol Surg 2002;28:619-22.
236. Cabrera J, Cabrera J Jr, García-Olmedo MA, Redondo P. Treatment of venous malformations with sclerosant in microfoam form. Arch Dermatol 2003;139:1409-16.
237. Donnelly LF, Bissett GS 3rd, Adams DM. Combined Sonographic and Fluoroscopic Guidance: a Modified Technique for Percutaneous Sclerosis of Low-Flow Vascular Malformations. Am J Roentgenol 1999;173,655-7.
238. Parsi K, Pereira J. Fluoroscopic ultrasound guided sclerotherapy. 16th International Workshop on Vascular Anomalies. International Society for the Study of Vascular Anomalies. Milan, Italy; 2006. p. 66-7.
239. van der Linden E, Overbosch J, Kroft LJ. Radiofrequency ablation for treatment of symptomatic lowflow vascular malformations after previous unsuccessful therapy. J Vasc Interv Radiol 2005;16:747-50.
240. Sidhu MK, Perkins JA, Shaw DW, Bittles MA, Andrews RT. Ultrasound-guided endovenous diode laser in the treatment of congenital venous malformations: preliminary experience. J Vasc Interv Radiol 2005;16:879-84.
241. Villavicencio JL. Surgical management of venous malformations. Commentary to D. Loose Paper. Venous Digest 2008.
242. Mattassi R. Surgical treatment and strategies in vascular malformations. Przeglad Flebologiczny 2005;13:145-9.
243. Mattassi R. Individual indications for surgical and combined treatment in so-called inoperable cases of congenital vascular defects. In: Balas P, editor. Progress in angiology 1991. Torino: Minerva Medica; 1992. p. 383-6.
244. Loose DA. Surgical management of venous malformations. Phlebology 2007;22:276-82.

Corresponding authors: B. B. Lee, M.D., PhD, F.A.C.S. Professor of Surgery and Director, Center for Vein, Lymphatics and Vascular Malformation, Georgetown University School of Medicine, Washington, D.C., USA.
E-mail: bblee38@comcast.net