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Radiologic Management of Vascular Malformations' Interventional, Classification and Diagnosis

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Abstract
This study aimed at analyzing the diverse group of congenital vascular malformations, with respect to their place within the broader classification of vascular anomalies and their pathologic, clinical, and radiologic diagnosis and management. And the study discuss some of the techniques, agents, and approaches used in the interventional treatment of this difficult group of lesions. The researchers are aware and acknowledge that there are several different techniques and agents that can be used to treat these lesions. The techniques and agents described in this article have been used for years by the experts with good results. The aim of this study is to share experience in the management of vascular malformations with these techniques at Jordanian hospitals, and to assess the patient satisfaction levels by the evaluation of the follow-up of patients with vascular malformations treated in the Interventional Radiology Unit from January 2016 to December 2016. Patients were classified according to the hemodynamics of the lesions (high- vs. low-flow).

1.1 Introduction

Vascular anomalies are prevalent in the general population and may produce significant impairment in quality of life. In recent years, minimally invasive interventional radiology techniques have become an efficient alternative in the treatment of these patients.

In late decades, an awesome arrangement has been found out about the histopathology, etiology, and treatment of vascular anomalies, which has caused change in the grouping and phrasing used to portray these injuries. The International Society for the Study of Vascular Anomalies (ISSVA) order framework, which has been broadly grasped by different subspecialists who nurture patients with these deformities, gives an approach in view of histopathology, clinical course, and treatment.

Be that as it may, utilization of more established terminology keeps on causing perplexity, off base judgments, and potential fumble. The all-encompassing objective of this article is 2-overlap. To begin with, to survey the ISSVA grouping of vascular oddities by examining key pathogenesis, imaging components, and current treatment of agent sores from every classification; and second, to talk about phrasing used to depict vascular contortions with the objective of clearing up dated or confounding terms that stay being used (Lowe, Marchant, Rivard & Scherbel, 2012).

Vascular anomalies constitute a normally experienced condition in the all-inclusive community. They may bring about corrective modifications, intermittent contaminations, practical debilitation, heart disappointment, seeping (among different disarranges), which convert into weakening of patients nature of life. The general order isolates them into tumors (hemangiomas) and vascular malformations. The characterization of vascular abnormalities has experienced numerous adjustments as of late, with a specific end goal to make it clear that hemangiomas and vascular malformations are distinctive pathologies; in this way, they require diverse medicines (Vargas, 2011).

Vascular malformations involve a gathering of injuries portrayed by the nearness of ordinary develop endothelial covering. These injuries are normally present during childbirth, in spite of the fact that not generally saw, and become equivalent with the kid's advancement. Jackson et al separated vascular malformations as indicated by their stream highlights (ie, low-and high-flow injuries). Low-stream vascular malformations include lymphatic, venous, and capillary malformations. The high-stream sores are arteriovenous malformations (AVMs)/fistulas (Dabus & Benenati, 2013).

Apart from capillary malformations, which are not usually treated with interventional therapy, all other types of vascular malformations can be treated with interventional techniques that typically require transarterial, transvenous, or direct access. Interventional treatment of vascular malformations has gained wider acceptance in recent years and is considered the first line of therapy in many centers. The results of interventional treatment vary depending on the type of vascular malformation being treated (Zheng, Zhou, Yang, Wang, Fan, Zhou & Suen, 2010).

Few areas within medical diagnosis are fraught with as many persistent misconceptions and misnomers as within the group of vascular anomalies.

Verifiably, morphologically assorted cutaneous and instinctive pigmentations, becomes flushed, masses,

and strange vascular spaces were named and sorted basically by net appearance, area, liquid substance, and an as often as possible covering and flighty clinical course.

Therefore, various repetitive, questionable, and notwithstanding deceptive terms have gathered in the course of the most recent 2 centuries depicting this arrangement of vascular imperfections. In everything except the biggest referral focuses, the issue of befuddling classification has exacerbated a general absence of demonstrative and restorative commonality with this gathering of elements and has brought about intrusive and frequently wrong treatment being established on alarmingly high rates of misdiagnosis (Mathur, Rana & Bothra, 2005).

There are several alternatives for treating these conditions, including the conservative dermatological-medical approach, the surgical management, and interventional radiology. The latter has made a significant advancement in recent years with the development of new embolic agents and the application of minimally invasive techniques. Transarterial embolization is generally considered the treatment of choice for high-flow vascular malformations. As for low-flow lesions, direct percutaneous puncture technique with embolic agents has proved successful (Do, Yakes & Shin, 2005).

1.2 Problem statement

This article focuses on the diverse group of congenital vascular malformations, with respect to their place within the broader classification of vascular anomalies and their pathologic, clinical, and radiologic diagnosis and management. It also we discuss some of the techniques, agents, and approaches used in the interventional treatment of this difficult group of lesions. The researchers are aware and acknowledge that there are several different techniques and agents that can be used to treat these lesions. The techniques and agents described in this article have been used for years by the experts with good results. The aim of this study is to share experience in the management of vascular malformations with these techniques at Jordanian hospitals, and to assess the patient satisfaction levels by the evaluation of the follow-up of patients with vascular malformations treated in the Interventional Radiology Unit from January 2016 to December 2016. Patients were classified according to the hemodynamics of the lesions (high- vs. low-flow). Complications and patient satisfaction with the treatment were assessed.

1.3 Objectives of the study

This study aimed at sharing the experience in the management of vascular malformations with the discussed treatment techniques at Jordanian hospitals, and also aimed at assessing the patient satisfaction levels by the evaluation of the follow-up of patients with vascular malformations treated in the Interventional Radiology Unit from January 2016 to December 2016. Patients were also classified according to the hemodynamics of the lesions (high and low-flow) and the complications and patient satisfaction with the treatment were assessed.

1.4 Methods and procedures

1.4.1 Classification of vascular anomalies

In 1982, Mulliken and Glowacki distributed a point of interest article proposing portrayal of vascular imperfections in view of biologic and pathologic contrasts. Their work separated between two noteworthy classes of vascular lesions: hemangiomas and vascular malformations. Hemangiomas were portrayed as injuries showing a past filled with quick neonatal development and moderate involution described by hyper cellularity amid the multiplying stage and fibrosis and decreased cellularity amid the involution stage. The term hemangioma, particularly in adults, is considered inaccurate and misleading in modern nomenclature and should be discarded. Vascular malformations were subdivided further into lesions consisting of capillary, venous, arterial, lymphatic, and initially fistulous networks as shown in table 1.

Table (1): Classification of vascular lesions in infants and children

Hemangiomas	Malformations
Proliferating phase	Capillary
Involution phase	Venous
	Arterial
	Lymphatic
	Fistulae

Source: Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. Plast Reconstr Surg 1982;69:413.

Expanding on introductory portrayal of angiographic stream designs by Burrows and partners, a corresponding arrangement conspire was proposed by Jackson and partners that considered flow rate as a variable deciding fitting examination and treatment. Vascular anomalies were isolated into low- flow venous malformations (VMs) and high- flow arteriovenous malformations (AVMs) with particular order for lymphatic malformations (LMs) and for hemangiomas.

Belov (1989) presented an etiologic and pathophysiologic grouping framework concentrated on the embryologic site of beginning of the deformity that prompted the advancement of every specific mutation. Every distortion sort was subdivided into two fundamental anatomic/pathologic structures: (1) truncular and (2) extratruncular. The truncular frame came about because of a generally late embryologic imperfection or occasion emerging inside a separated vascular trunk. This form is often more severe and classified as due to a vascular aplasia, obstructive, or dilatory phenomenon. The extratruncular shape, regularly less extreme, emerged because of a moderately early embryonal dysplasia inside the primitive undifferentiated narrow system and could display in a diffuse/invading or constrained/confined mold, as appeared in table 2.

Table 2): Anatomopathologic classification of vascular defects
(Hamburg classification)

Type	Forms	
	Truncular	Extratruncular
Predominantly arterial defects	Aplasia or obstructive Dilation	Infiltrating Limited
Predominantly venous defects	Aplasia or obstructive Dilation	Infiltrating Limited
Predominantly lymphatic defects	Aplasia or obstructive Dilation	Infiltrating Limited
Predominantly asteriovenous shunting defects	Deep Superficial	Infiltrating Limited
Combined/mixed vascular defect	Arterial and venous Hemolymphatic	Infiltrating Hemolymphatic Limited Hemolymphatic

1.5 Study sample

Eighty-nine patients were included in the this study, 41 males and 48 females, as shown in table 3. Mean age was 21.5 years. When analysis of results was carried out, 67 patients had already been discharged and required no more sessions of minimally invasive treatment. The remaining 22 patients were still under medical observation, with ongoing treatment sessions. Twenty-six patients (29.2%) had received various treatments prior to interventional radiology; incisional or excisional biopsies; OK-432 therapy (Picibanil); use of carbon dioxide snow, and/or sclerosis by bleomycin.

Table (3): Characteristics of the study sample

Mean age		21.5 years
Sex	Male	41
	Female	48
Patients still under medical observation		22 (24.72%)
High flow		12 (13.5%)
Low flow		10 (11.23%)
Discharged patients		67 (75.3%)
High flow		16 (17.8%)
Low flow		51 (57.3%)

In over 55 % of patients, the vascular anomaly was located either on the face or on lower extremities as shown in table 4.

Table (4): Location of vascular malformations

Location	Number of patients	Percentage
Head and neck	35	39.32%
Low limbs	33	37.1%
Pelvic region	5	5.6%
Neck	6	6.4%
Upper limbs	5	5.6%
Chest	3	3.3%
Lumbar region	2	2.2%

Out of the 89 patients, 27 (30.3%) had high flow malformation (pure and mixed), and 61 (68.5%) exhibited low-flow lesions (vascular malformations, lymphangiomas, or mixed). The detail is shown in table 5, and figs. 1).

Table (5): Types of vascular malformations

Vascular malformations	Frequency	Percentage
Low flow	**61**	**68.5%**
Low flow	49	55%
Lymphangioma	8	8.9%
Lymphangioma + low flow	3	3.37%
Low flow AVF	1	1.12%
High flow	**27**	**30.3%**
High flow	22	24.7%
Lymphagioma + high flow	5	5.62%

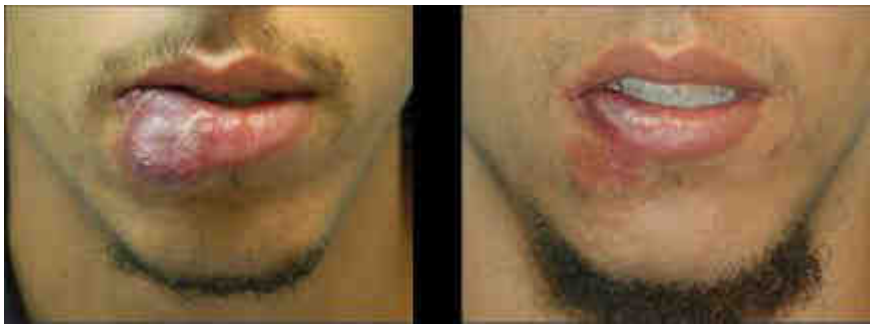

Fig. (1): Patient with high-flow malformation, pre- and post-treatment.

1.6 Clinical diagnosis

The diagnosis of essence and kind of vascular malformation typically can be made absolutely on clinical history and physical examination. By definition, every vascular malformations are available during childhood, and most end up noticeably obvious to some degree amid early stages or adolescence. Injuries named as " acquired" amid pre-adulthood are generally those of deficient size or indications to have been identified amid the time of pari passu development in youth, which turned out to be clinically apparent as the sore experienced proceeded with straight development after typical physical development stopped. Vascular malformations never relapse or involute and may develop at a rate more noteworthy than ordinary substantial development. Patients' complaints are often a result of localized swelling from exertional or postural venous stasis and paroxysmal localized thrombosis causing pain, and compression or dysfunction of adjacent muscle and nerves. Aside from cosmetic factors, lesion size, location, and proximity to crucial structures dictate the nature and severity of patient symptoms (Cappabianca, Cinalli, Gangemi, Brunori, Cavallo, Divitiis & Godano, 2008).

AVMs are a distinct entity characterized by high-flow physiology and an aggressive clinical course. The lesion can be detected at birth in 40% of cases and most commonly occurs in the extremities and pelvis. The Schobinger clinical staging system, introduced at the 1990 meeting of the International Workshop for the Study of Vascular Anomalies in Amsterdam, outlines the inevitable clinical course of untreated AVMs from their local skin and soft tissue effects to their eventual systemic cardiovascular compromise (Temple & Marshalleck, 2014), as shown in table 6.

Table (6): Schobinger clinical staging system of arteriovenous malformations

Stage one	Quiescence	Cutaneous blush, skin warmth, arteriovenous shunt on Doppler ultrasound
Stage two	Expansion	Darkening blush, lesion shows pulsation, thrill and bruit
Stage three	Destruction	Steal, distal ischemia, pain, dystrophic skin changes, ulceration, necrosis, soft tissue and bony changes
Stage four	Decompensation	High-output cardiac failure

1.7 Imaging diagnosis of vascular malformations

Although clinical history and examination are sufficient to establish the diagnosis of a vascular malformation, imaging is an indispensable part of the full patient workup. In addition to confirming the diagnosis, defining the extent of the lesion, and detecting often occult associated pathologic findings, imaging allows feasibility assessment and planning of any potential percutaneous image guided or surgical therapy.

Conventional radiography: Because of inherent low soft tissue contrast resolution, conventional radiography provides little direct information on the lesion in question other than its mass effect and only if of sufficient size. Dystrophic calcification, either diffuse or in the form of phleboliths, can be identified within VMs. Of vascular malformations, 34% cause some form of adjacent bony change. In the extremities, VMs are more likely to cause bony hypoplasia and demineralization; however, the authors commonly have noted local mixed sclerotic change and periosteal reaction within adjacent bone. LMs can cause hypertrophy and bony distortion, whereas AVMs are more likely to cause destructive and intraosseous change or skeletal overgrowth.

Ultrasonography: Ultrasound evaluation is noninvasive, inexpensive, and readily available. In addition to gray-scale characterization of the lesion morphology and defining its size and extent, Doppler evaluation is invaluable in the discrimination between high-flow and low-flow malformations. On gray-scale imaging, VMs are compressible; show heterogeneous echotexture (98%); and appear hypoechoic (82%), isoechoic (8%), or hyperechoic (10%) relative to adjacent subcutaneous tissue. Phleboliths with acoustic shadowing are highly specific for VM, but are seen in only 16%, and only a few show discernible anechoic vascular spaces. LMs have a variable appearance, with macrocystic LMs showing septa separating anechoic cavities that can contain debris Microcystic LMs have small cavities resulting in innumerable reflective interfaces and a hyperechoic appearance. AVMs have a heterogeneous echotexture and reveal large tubular vascular structures without a well-defined soft tissue mass, as shown in fig. 2(A,B).

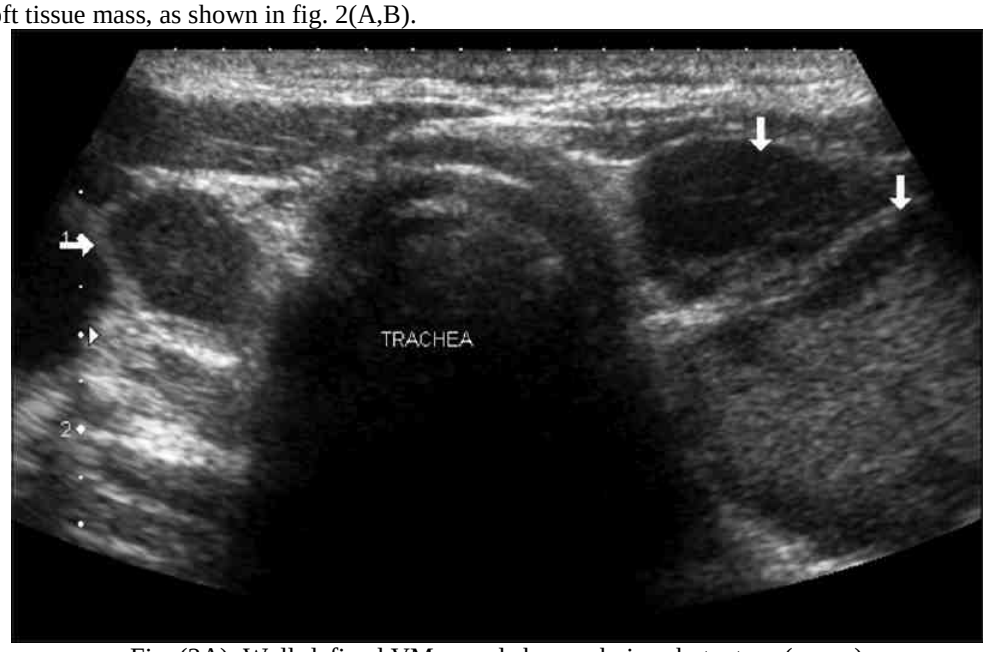

Fig. (2A): Well-defined VM reveals hypoechoic echotexture (arrow)

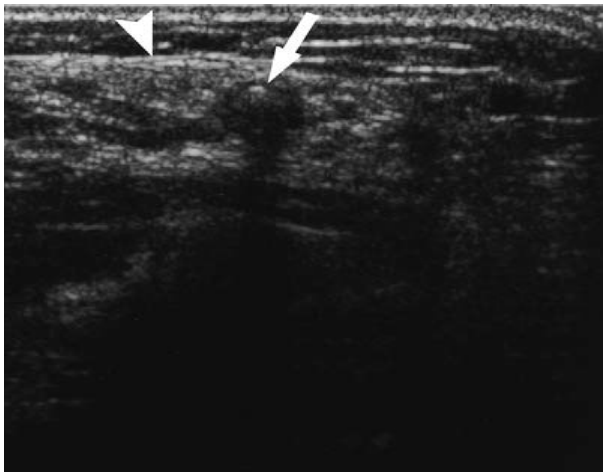

Fig. (2B): VM appears ill defined and mildly hyperechoic (arrowhead)

Color Doppler and pulsed Doppler interrogation of VMs reveals monophasic flow in 78%, biphasic flow in 6%, and no flow in 16%. Biphasic flow may be characteristic of mixed lesions, and the absence of flow may represent lesion thrombosis versus flow below detectable limits. LMs do not show detectable flow on Doppler; however, flow can be detected within lymphatic cyst wall and intervening tissues. AVMs have high vessel density, high systolic flow, arteriovenous shunting, and arterial flow
within enlarged draining veins, as shown in fig. 3 (A,B,C).

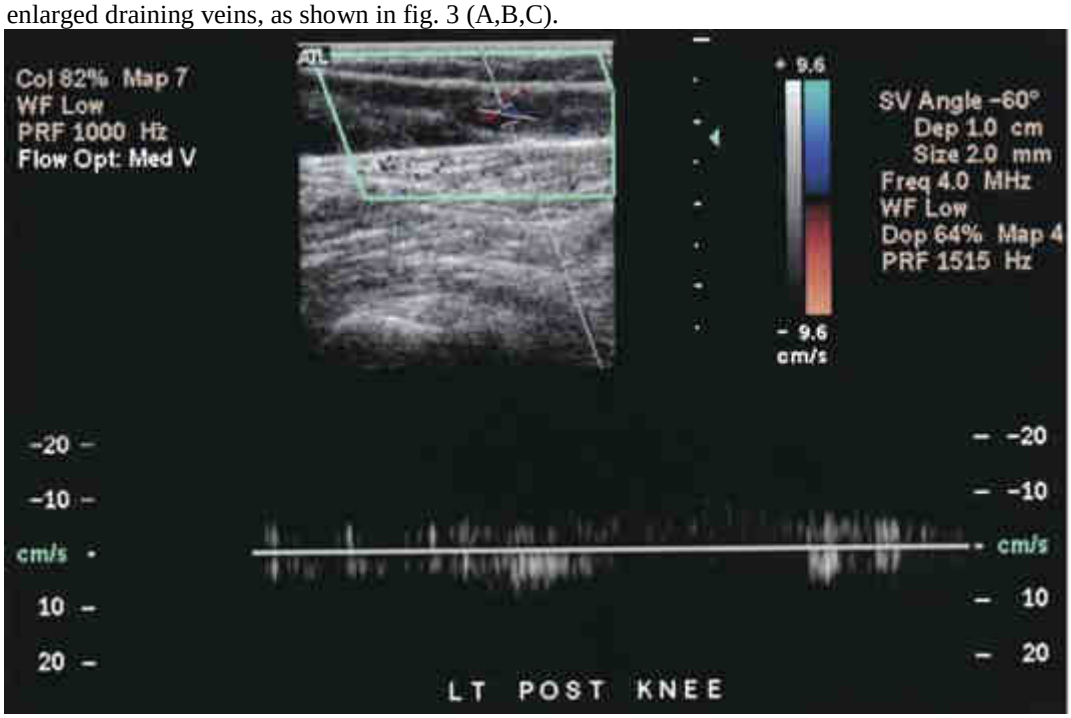

Fig. (3A): VM reveals mixed venous waveform

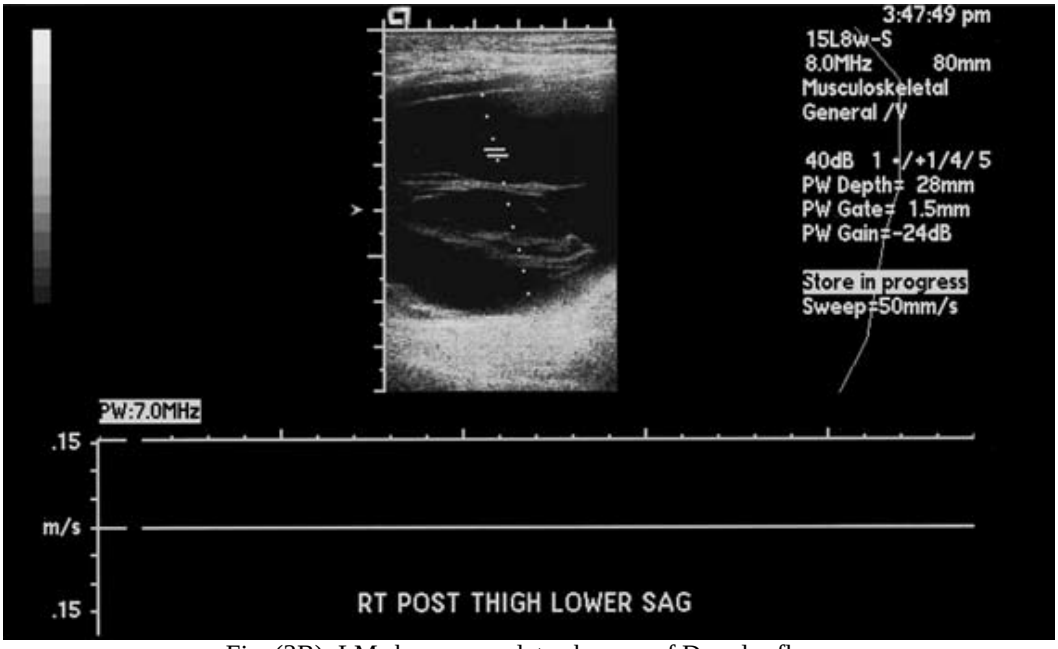

Fig. (3B): LM shows complete absence of Doppler flow.

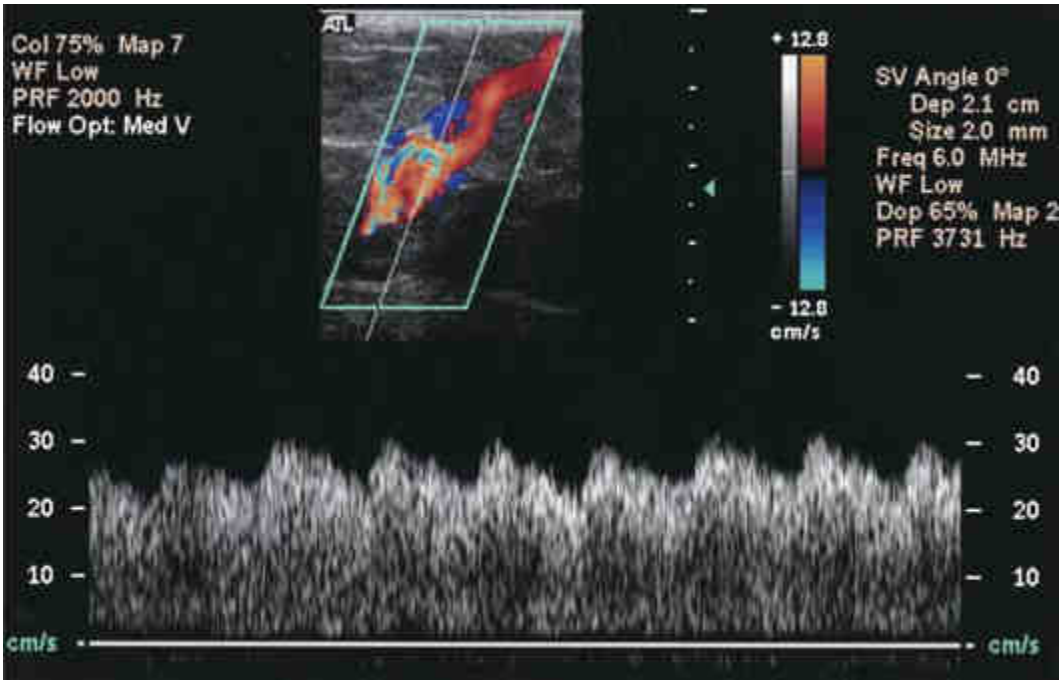

Fig. (3C): VM reveals multidirectional flow and high-amplitude arterial waveform with spectral broadening

1.8 Therapeutic options for vascular malformations

Essential to the optimal care of patients with vascular malformations is the assembly of an experienced multidisciplinary team that is well versed in the latest diagnostic and therapeutic techniques and controversies within the field of study of vascular anomalies. This level of familiarity and expertise usually can be achieved only through frequent exposure to vascular anomalies facilitated through streamlined interdisciplinary communication and usually is found at larger referral institutions in the form of a vascular anomalies center. Based on the specific patient population, such a team comprises many specialties, including dermatology, vascular or plastic and reconstructive surgery, otolaryngology, orthopedic oncology, anesthesiology, pediatrics, radiology, and rehabilitation medicine supported by physiotherapy or occupational therapy.

One should arrive at the decision to treat a vascular malformation by consensus between referring specialties, with careful consideration of the potential procedure's associated morbidity relative to the present and often uncertain future morbidity if left untreated. Lee devised a ''decision to treat'' formula in which therapy

is initiated based on the patient possessing at least one absolute or two relative indications. Absolute indications include hemorrhage; progressive high output failure; complications secondary to venous hypertension; and lesion location within a life-threatening area, such as the airway, or lesion location threatening vital functions. Abbreviated relative indications include progressive disabling pain or discomfort, functional disability or impairment affecting daily life and quality of life, cosmetically severe deformity, vascular-bone syndrome causing growth discrepancy, lesion location at high risk for complication, and recurrent infection or sepsis.

In the case of flow malformations, where appropriate, invasive management should be reserved for patients who meet the above-mentioned criteria after failing more minimal therapies.

VMs often can be treated at least initially successfully with elevation, compression garments, and aspirin, whereas medical management of LMs requires antibiotics and steroids during infectious or hemorrhagic episodes.

The decision of whether to treat vascular malformations via surgery or interventional radiologic techniques is complex and often dictated by factors specific to the lesion in question, patient preference, and availability of expertise or patterns of practice within a given institution. Several attempts have been made to define more clearly treatment modality algorithms between surgery and interventional radiology based on lesion type, size, location, and morphology.

Overall, the Hamburg classification of vascular malformations provides a framework through which general trends in current therapy can be observed. Truncular VMs, often more extensive and requiring correction of hemodynamic factors, are treated primarily surgically with or without adjunctive radiologic embolosclerotherapy. There is little role for radiologic management of other truncular lesions. Conversely, extratruncular forms of VMs and LMs incorporate interventional radiologic therapy to a much greater degree. AVMs are largely treated via percutaneous means with or without adjunctive surgical resection.

1.9 Conclusion

Confusion still exists as to the classification and nomenclature of vascular anomalies, which has an impact on the clinical diagnosis and management. The term hemangioma should be reserved for lesions histologically exhibiting rapid hypercellular endothelial growth, most commonly seen in the form of infantile hemangioma in pediatric patients. Essentially all other vascular anomalies are termed vascular malformations and are described as being present at birth and growing commensurately or pari passu with the child.

Vascular malformations are characterized by a normal rate of endothelial cell turnover comprising vascular channels lined with flat “mature” endothelium. Vascular malformations can be divided into venous, lymphatic, arteriovenous, or mixed varieties. These can be divided further into truncular lesions, which result from a relatively late embryologic defect or event arising within a differentiated vascular trunk, or extratruncular lesions, which result from a relatively early embryonal dysplasia within the primitive undifferentiated capillary network during vasculogenesis and angiogenesis.

These latter two processes occur by interaction between primitive endothelial cells and the adjacent embryonic mesenchyme to produced ordered vascular channels containing smooth muscle and adventitia. A molecular defect in this process is hypothesized as the cause of many vascular malformations.

VMs and LMs constitute the low-flow category and can have a varied appearance, may be solitary or multiple, may be localized or infiltrating, and can occur anywhere in the body. Patients present with functional disability and compression of vital structures or pain secondary to mass effect and in the case of VMs pain owing to stasis and thrombosis. LMs are divided further into macrocystic, microcystic, and mixed varieties. AVMs constitute the high-flow category and consist of a nidus of abnormally developed arteriolar venular interface without a normal intervening capillary network. They can occur anywhere and follow a predictable clinical course according to the Schobinger classification, with locally aggressive symptoms of pain, congestion, and erythema progressing to high output failure.

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