Vascular Malformation, General

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There has been nosological confusion among various medical specialists involved in the management of superficial vascular anomalies. In the past, common disease names for different descriptions have been used to define these diseases. The international Society for the Study of Vascular Anomalies (ISSVA) was born in 1992. A very basic classification system was adopted by the ISSVA during its 1996 workshop[1,2], which has become accepted and used worldwide. Purpose of ISSVA classification is to lead appropriate clinical diagnosis and treatment strategy with differentiation of vascular tumors (hemangiomas) and vascular malformations using simple and easily comprehensible nomenclature.

This system is based on the founding biological investigation of Mulliken and Glowacki published in 1982, which provided the groundwork for a proper identification of vascular birthmarks[3]. Vascular tumors have been differentiated from vascular malformations based on their clinical appearance, radiological and pathological features, and biological behavior.

Vascular tumors grow by cellular (mainly endothelial) hyperplasia: the very common infantile hemangioma is in reality a benign vascular tumor. In contrast, vascular malformations have a quiescent endothelium and are considered to be localized defect of vascular morphogenesis, likely caused by dysfunction in pathways regulating embryogenesis and vasculogenesis. Infantile hemangiomas grow rapidly after birth and then involute gradually in childhood. Hence, generally they don't require aggressive treatments, except for lesions with serious functional disorder. Vascular malformations never regress and persist throughout life. Most of them have commensurate growth during childhood, and some worsen over time if not treated. They cause symptoms including pain, ulcer, hemorrhage, infection, overgrowth of limb, functional disorder, cosmetic problems and so on, which require aggressive treatments, such as embolization, sclerotherapy, or surgical resection[2].

A subdivision of vascular malformations, based on hemodynamics and on predominant anomalous channels, was created. Vascular malformations are either slow-flow or fast-flow, and they are subcategorized into capillary malformation (CM), venous malformation (VM), lymphatic malformation (LM) and arteriovenous malformation (AVM). Some patients have complex-combined vascular malformations.

In the international workshop of ISSVA held in Melbourne in 2014, an updated ISSVA classification was presented and accepted **(Table 1)** [4,5,6]. The new updated version, distinguishes CM, LM, VM, AVM and arteriovenous fistulas (AVF). Also defects of the main named vessels, combined forms, and association with other anomalies are considered. Several subgroups will be included in order to include all new discoveries, like the genetic-related clinical pictures and others[6].

This presentation will focus on ISSVA classification and general clinical information regarding especially infantile hemangioma **(Fig.1)** and three types of vascular malformation, including VM **(Fig. 2)**, AVM **(Fig. 3)**[7], and LM **(Fig 4)**. Treatments for the various vascular anomalies have become more specifically adapted over last 30 years. Surgical procedures have been adapted and customized by both plastic and vascular surgeons. Therapeutic embolization and sclerotherapy now have clear indication for use. A great deal of progress has been achieved in the field of vascular anomalies, in particular to improve our knowledge of their pathogenesis and the results of therapy[6].

Vascular anomalies				
Vascular tumors	**Vascular malformations**			
	Simple	**Combined**	**of major named vessels**	**associated with other anomalies**
Benign	CM	CVM	Affect	Klippel-Trenaunay syndrome
Infantile hemangioma	LM	CLM	lymphatics	Parkes Weber syndrome
Congenital hemangioma	VM	LVM	veins	Servelle-Martorell syndrome
etc.	AVM	CVLM	arteries	Sturge-Weber syndrome
Locally aggressive	AVF	CAVM		Limb CM + congenital non-progressive
or borderline		CLAVM	Anomalies of	limb hypertorophy
Kaposiform		others	origin	Mafficci syndrome
hemangioendothelioma			course	Macrocephaly-CM
etc.			number	Microcephaly-CM
Malignant			length	CLOVES syndrome
Angiosarcoma			diameter	Proteus syndrome
etc.			valves	Bannayan-Riley-Ruvalcaba syndrome
			communication	
			persistence	

Table 1: Updated ISSVA classicication (last revision May 2018) [4]

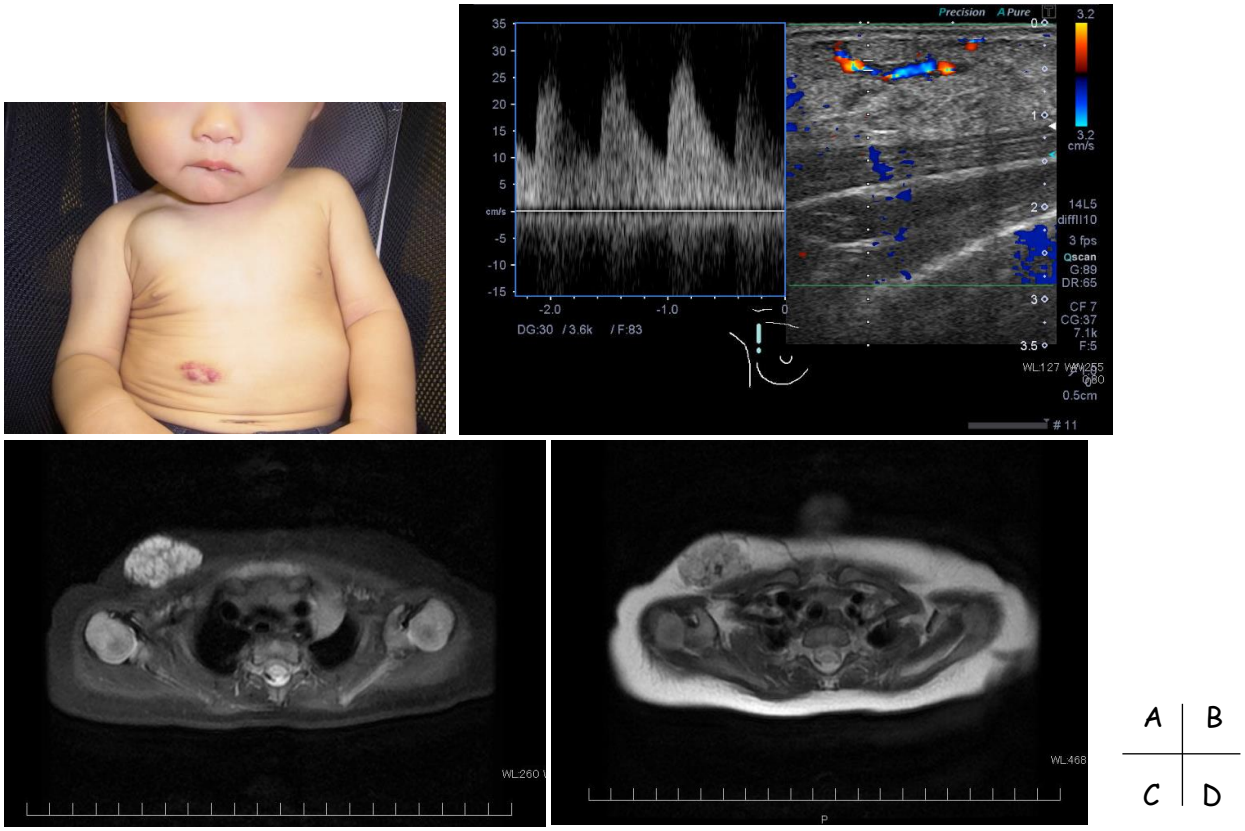
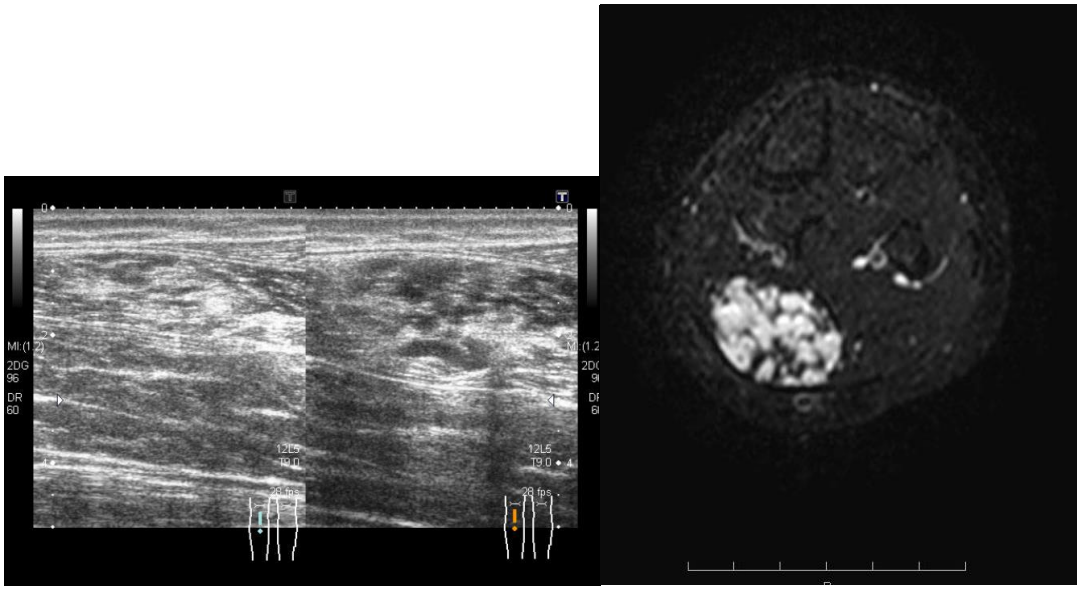
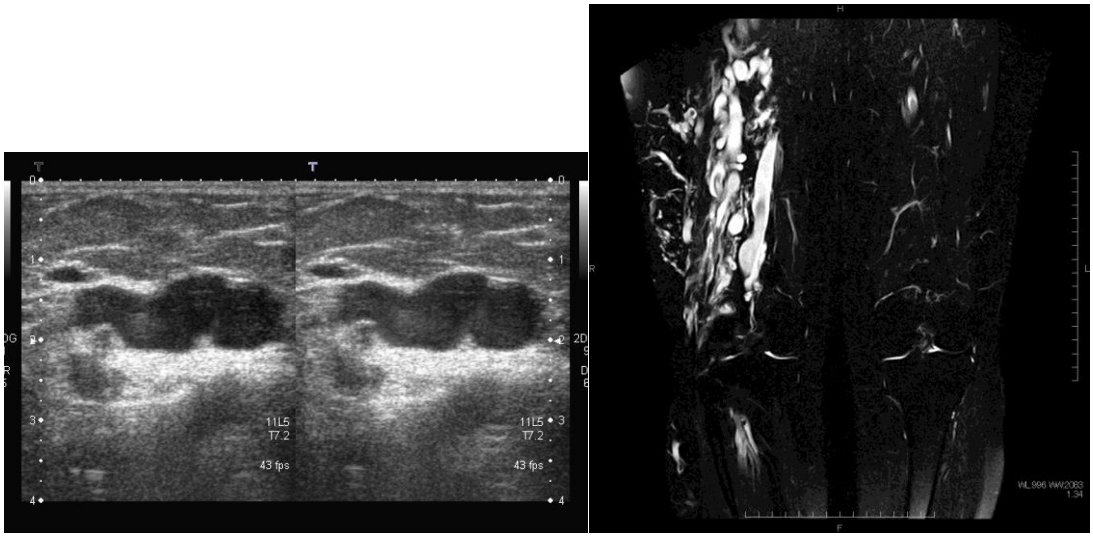


Figure 1: Infantile hemangioma of the chest wall.

A: Photograph. B: Ultrasound. C: MRI axial T2WI. D: MRI axial STIR.



A1 A2



B1 B2

Figure 2: Venous malformation.

A: Lobular type venous malformation of the lower leg. 1: Ultrasound. 2: MRI axial STIR.

B: Varicose type venous malformation of the thigh. 1: Ultrasound. 2: MRI coronal STIR.

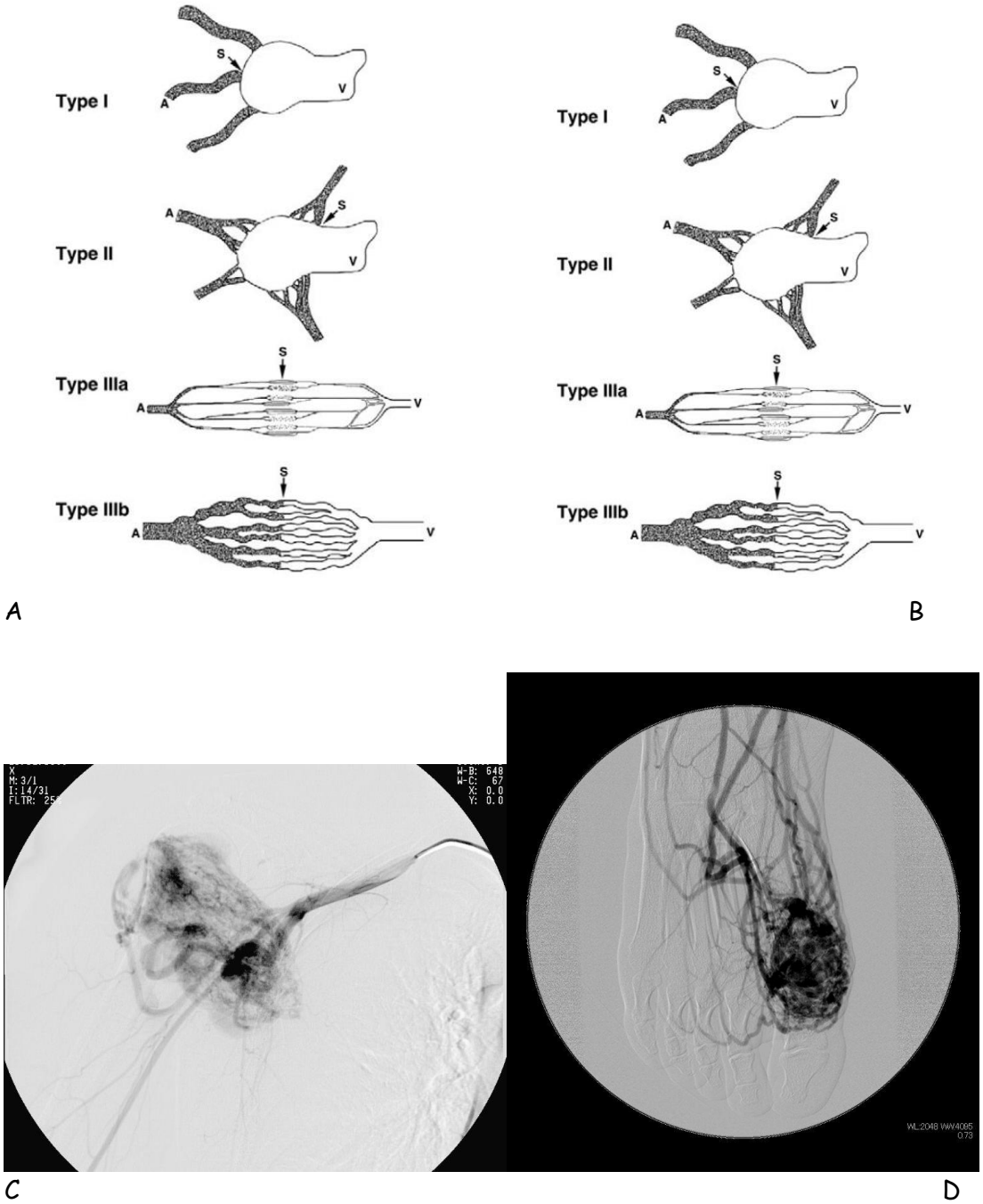


Figure 3: Arteriovenous malformation.

A: Angiographic classification of peripheral arteriovenous malformations[7].

B: Type II arteriovenous malformation of the elbow. Angiography.

C: Type IIIa arteriovenous malformation of the shoulder. Angiography.

D: Type IIIb arteriovenous malformation of the foot. Angiography.

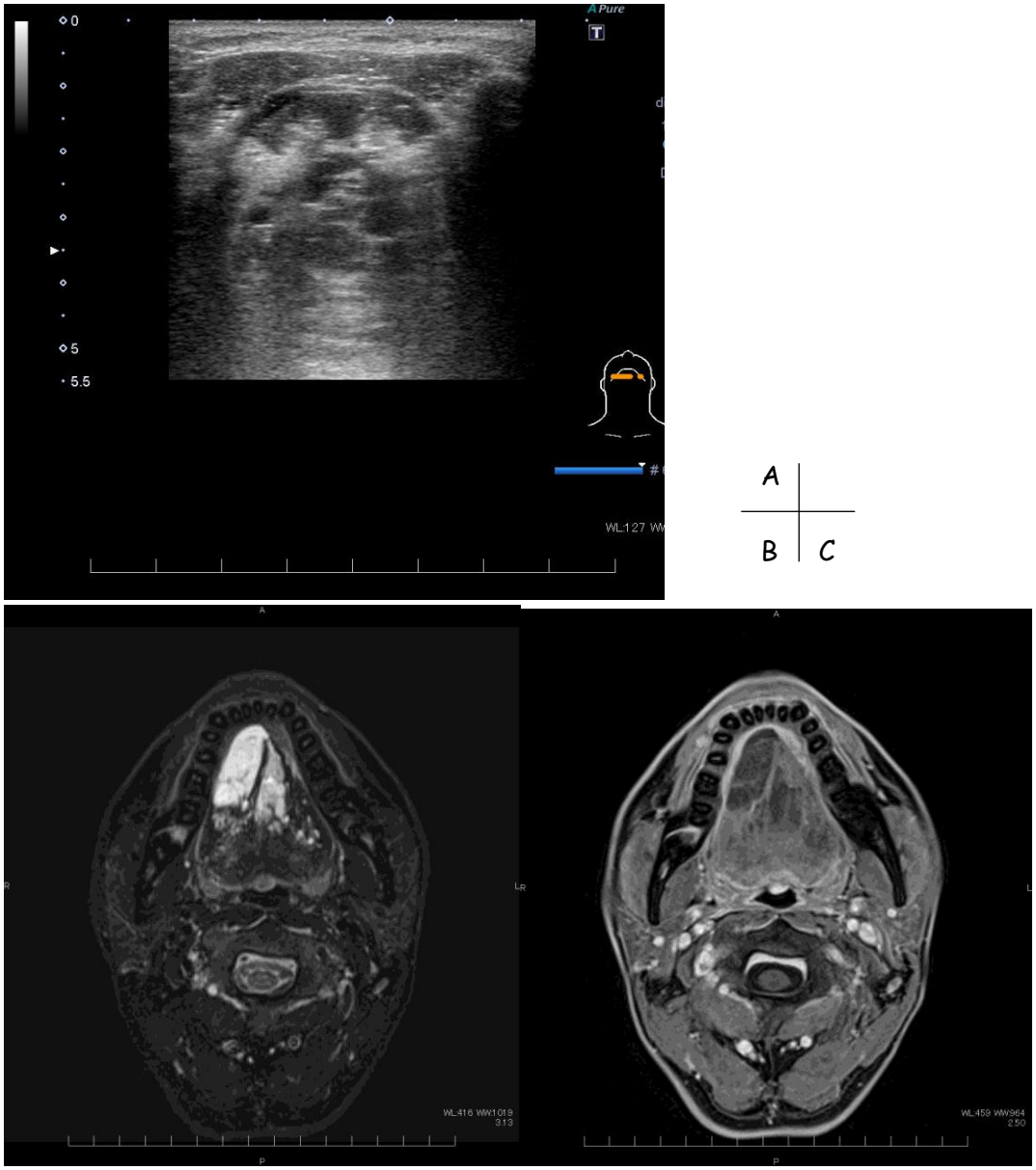


Figure 4: Lymphatic malformation of the tongue. Angiography.

A: Ultrasound. B: MRI axial STIR.

C: MRI axial fat-suppressed contrast-enhanced T1WI.

References

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