

Diagnosis and Treatment of Vascular Malformations

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ABSTRACT

Vascular malformations are complex lesions of the vasculature and may be congenital or acquired. The clinical signs of congenital malformations may be subtle in neonates, infants and young children and they usually become apparent during later childhood and adolescence. Acquired vascular malformations consist of arteriovenous fistulas and can be the result of trauma, including iatrogenic one. Vascular malformations include capillary, lymphatic, venous, arterial and combined malformations. Because of their great variability in type, site, extension and secondary effects, congenital vascular

malformations are difficult to understand and this can lead to diagnostic confusion and uncertainty. Treatment strategies include endovascular therapy, surgery, laser and medication depending on the symptoms, location and functional status of the lesion. The aim of therapy is to reduce severe symptoms or complications of the lesion. In this review we use a systematic approach for the vascular malformations, based on the clinical aspects, haemodynamic characteristics and morphology of the lesions with emphasis on the endovascular treatment options.



KEY WORDS

vascular malformations; sclerotherapy; arteriovenous malformations; embolisation



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Introduction

Vascular anomalies are structural irregularities of the vasculature, which occur during vasculogenesis, angiogenesis and lymphangiogenesis [1].

These are the major processes that give rise to, and maintain, the adult lymphatic and vascular systems. Vascular anomalies include vascular tumours (which consist mainly of infantile haemangiomas) and vascular malformations that are divided according to the component on which they arise. These include capillary, lymphatic, venous, arterial and combined malformations. Because of their great variability in type, site, extension and secondary effects, congenital vascular malformations (CVMs) are difficult to understand and this can lead to diagnostic confusion and uncertainty.

CVMs are always present at birth, but the clinical signs may be subtle in neonates, infants and young children [2].

The lesions enlarge in time proportionately to child growth and hence the physical signs evolve over time. They usually become symptomatic during later childhood and adolescence and they do not regress spontaneously [3]. The aim of therapy is to reduce severe symptoms or complications of the lesion. Erroneous or incomplete therapy may lead to worsening of symptoms and morbidity. Therefore, treatment strategy should be individualised and carefully planned, based on a multidisciplinary approach with full integration of endovascular treatment and, in some cases, open surgery to improve treatment outcome. In many cases the combination of endovascular treatment and open surgery is necessary for the best possible outcome [4].

In this review we use a systematic approach for the vascular malformations, based on the clinical aspects, haemodynamic characteristics and the morphology of the lesions with emphasis on the endovascular treatment options.

Classification of Congenital Vascular Malformations

Few diseases have generated such a large number of different names as CVMs. Based on haemodynamics, we divide CVMs into low-flow malformations-AVMs (capillary, lymphatic and venous) and into high-flow lesions (arteriovenous malformations and arteriovenous fistulas-AVFs.), adding syndromes of combined complex cases [5]. Treatment strategy is mainly based on the haemodynamics of the lesion (**Table 1**) [6]. Every vascular



Fig. 1. Typical appearance of low-flow venous malformations presenting as blue soft compressible masses in the back (**a**) and the cheek (**b**) in two paediatric patients.

malformation with an arterial component can be called a “high-flow” malformation, whereas every vascular malformation without an arterial component is a “low-flow” malformation [7]. In the international workshop of ISSVA (International Society for the Study of Vascular Anomalies) held in Melbourne in 2014, an updated clas-

Table 1. Treatment strategy according to the haemodynamics of the lesion

High-flow lesions	Arteriovenous malformation	Embolisation (transarterial, transvenous, percutaneous) Surgery (of the embolised nidus)
	AVF	Embolisation (transarterial, transvenous)
Low-flow lesions	Venous malformations	Sclerotherapy Laser Surgery (by persisting lesions)
	Lymphatic malformations	Sclerotherapy (macrocytic type) Surgery Laser Immunomodulation therapy
	Capillary malformations	Laser treatment

sification was presented and officially accepted. The new classification, revised on May 2018, aims to include all subgroups of vascular defects, recent genetic data and appropriate therapy (www.issva.org).

Low-flow Malformations

Venous Malformations

Clinical presentation

Venous malformations (VMs) are present as blue, soft, partially or fully compressible masses, and are often blanchable on light pressure (**Fig. 1**) [8]. The enlargement may be positional, as lesions may fill up and enlarge in some positions or after exercise. The symptoms depend on the location and extension of the lesion. The patients may suffer from swelling and pain, usually due to thrombophlebitis and phleboliths. VMs involving the lower limb veins can cause venous hypertension. When located in the neck and facial area, venous malformations may lead to airway narrowing or symptoms from the orbital cavity.

Imaging characteristics

Ultrasound (US) examination is the ideal way to complete a clinical examination. In US a low-flow pattern is seen in echofree structures. Flow may be spontaneous or only evoked by provocation maneuvers. However, magnetic resonance imaging (MRI) is the best method to evaluate the extension of the malformation and its relationship with adjacent structures. The most common pattern of VMs is a cavitory lesion with a delayed enhancement with or without abnormal draining

vein. The first acquisition has to be performed with a wide field of view to assess the extent of the lesion. On T1-weighted images, a VM is hypointense or isointense compared to the muscle. Absence of flow-void is mandatory for the diagnosis of VM. On fat-sat T2-weighted sequences, high-signal intensity is observed. T2-weighted gradient-echo sequences can also be used to identify calcifications or haemosiderin. Heterogenous enhancement is seen after injection of gadolinium (**Fig. 2**) [9].

Therapy indications

Indication for therapy:

1. Pain and discomfort, related with thrombophlebitis or congestion
2. Compression and deformity of adjacent structures
3. Thromboembolic complications
4. Cosmetic deformities and functional disability

The goal of therapy is to treat the symptoms of the VM. The reduction of the VM's volume can relieve pain and swelling of the lesion, reduce the risk of coagulopathy and compression symptoms.

Treatment strategies

Sclerotherapy: Percutaneous sclerotherapy (**Fig. 3**) is the treatment of choice for these lesions. It is safer and more effective than surgery [10]. The sclerosing agents produce inflammation, thrombosis and cellular necrosis, which leads to shrinking of the lesion. A good clinical outcome with subjective improvement of symptoms (pain, discomfort) and simultaneously objective evidence of improved clinical signs (reduction of the size

of the lesion) can be achieved in 75-90% of the patients [11,12].

Technique of classic sclerotherapy: Sclerotherapy (**Fig. 4**) is usually performed under general anaesthesia, especially when aethanol is used as the sclerosing agent, because of the pain provoked during injection. The common approach is percutaneous direct puncture of the lesion. Preoperative MRI is helpful in planning the best approach to puncture the lesion. A small-bore needle is inserted percutaneously into the lesion usually using US guidance. After slow venous return is seen through the needle, a direct venography should be performed before injecting the sclerosing agent to confirm that the needle is within the lesion, to determine the size of the malformation and ensure that there is no extravasation into normal tissues and no filling of the deep veins. Direct pressure or use of a tourniquet to control outflow can be used. After determining the contrast volume necessary to fill the abnormal venous compartment, the same volume of sclerosing agent is infused slowly. In cases when there is concern regarding drainage of the lesion, the infusion is performed using the negative subtraction technique. It is advisable to perform the puncture of the lesion from multiple accesses and inject small doses of sclerosing agent at each point, in order to reduce the risk of local complications. Shrinkage of VMs usually begins 1-2 weeks after the procedure. After 6-8 weeks, if the desired degree of involution has not occurred, the procedure can be repeated.

Common sclerosing agents are highly concentrated alcohol, sodium tetradecyl sulfate (STS), polidocanol 3% and alcohol gel (Sclerogel®).

Absolute aethanol is the most effective sclero-agent. It is the only sclerosant that can induce denaturation of tissue protein and avoid regeneration of endothelial cells with subsequent permanent obliteration of the vessel lumen. The negative aspect of aethanol is its high toxicity, associated with local and systemic complications, such as local necrosis, hypoglycaemia and arrhythmia [13].

STS 3% (Sotradecol®, Fibro-Vein®) has been widely used as a sclerosing agent since 1946. STS is very effective and, together with polidocanol, the most commonly used sclerosing agent in the treatment of small varicose veins of the legs, as well as venous malformations,



Fig. 2. Low-flow, venous malformations of the cheek. **a.** Axial fat-sat T2-weighted image demonstrates a well-defined hyperintense lesion without perilesional oedema. Small signal voids (phleboliths) are detected within the lesion. **b.** Axial T1-weighted image shows an isointense lesion of the right cheek (arrows). **c.** Heterogeneous filling of the malformation by gadolinium is observed (arrows).

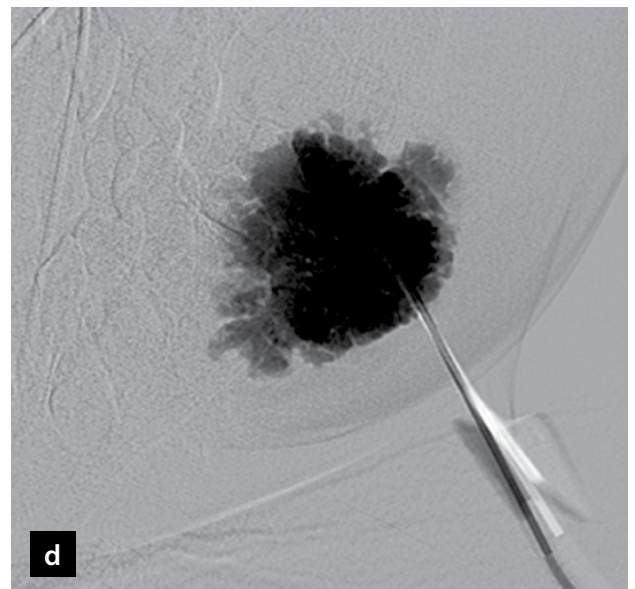
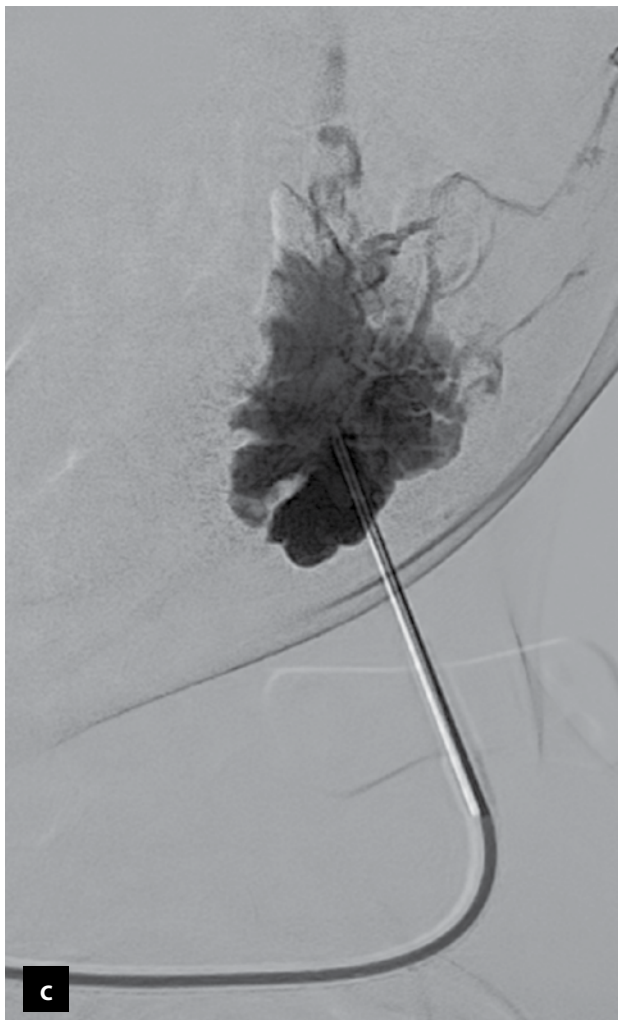
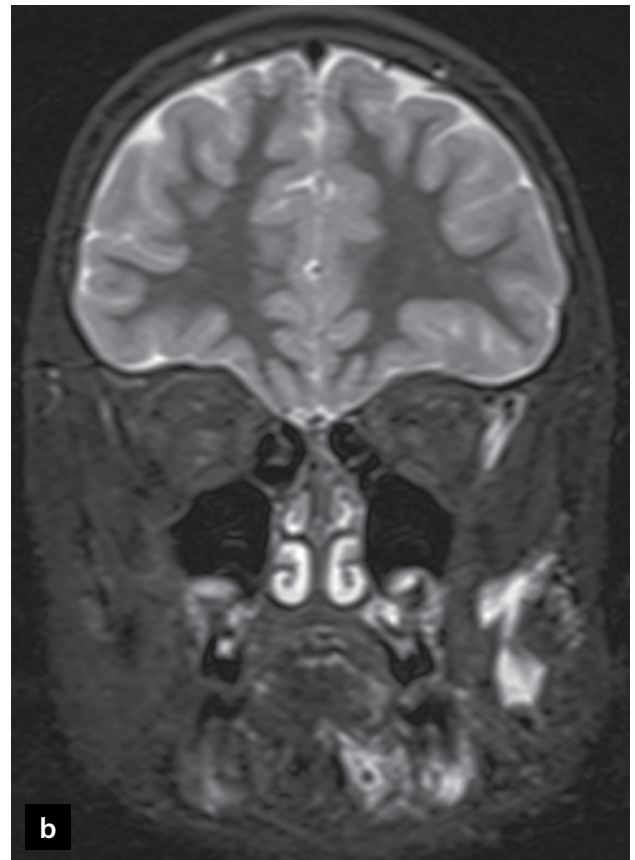
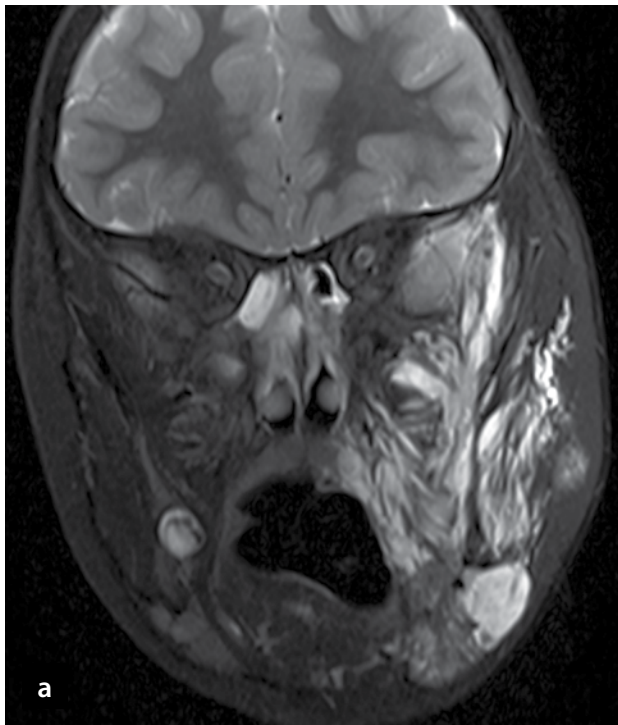


Fig. 3. Extensive low-flow venous malformation on the left side of the face, seen as diffuse lesion of high-intensity on fat-sat T2-weighted images before (a) and after (b) several treatments with percutaneous sclerotherapy. Direct percutaneous sclerotherapy illustrates a spongy pattern of slow-flow venous malformations (c, d).

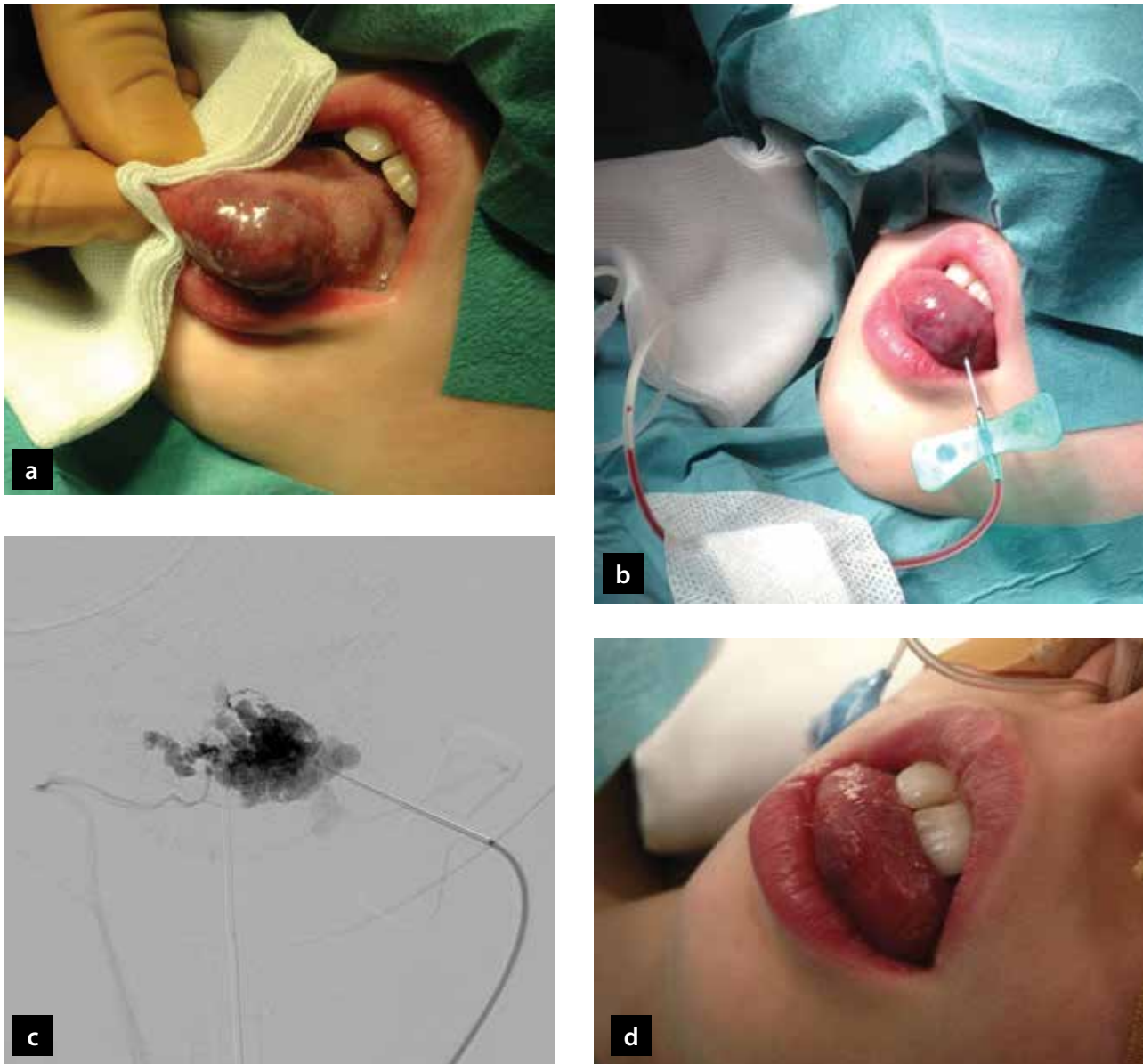


Fig.4. A 7-year-old female with low-flow venous malformation of the tongue. Before (a) during (b, c) and two months after (d) treatment with endovascular sclerotherapy in one session.

due to fewer complications than absolute aethanol. The mechanism of action of STS is to produce maximum endothelial damage with minimal thrombus formation, leading to fibrosis of the lesion which eventually shrinks. The vascular luminal obliteration may or may not be permanent [14].

Sclerogel® is also an effective and commonly used sclero-agent. It is a combination of jellied alcohol and a specific cellulose derivate, which reduces the risk of necrosis. Significantly less alcohol is needed per treatment, it is safer than absolute aethanol and easy to use.

Lymphatic malformations

Clinical presentation

Lymphatic malformations (LMs) present as a soft, partially compressible and extensive mass in the head and neck region. Patients may have advanced signs of lymphoedema of the limbs at a very young age. These include cutaneous changes such as erythema, induration, verrucous change and hyperkeratosis [15]. LMs are divided into macrocystic (cyst diameter >0.5 cm), microcystic (cysts <0.5 cm) and a combination of the two. Macrocystic LMs most commonly occur in the loose

connective tissue of the neck, axilla, chest wall or groin and often change in size.

The most common complications of LMs are bleeding and recurrent infections, that occur because of the impaired lymphatic drain, or cellulitis. Infections are seen in 71% of the patients and can lead to sepsis [16].

Imaging characteristics

In pure LMs, a no-flow pattern occurs in US examination. MRI exams demonstrate shape, extension and variety, macrocystic or microcystic (honeycombed lesions). MR imaging shows a septated mass with low-signal intensity on T1- and high-signal intensity on T2-weighted images. Because of varying amounts of protein or haemorrhage within the lesion, LMs occasionally show variable signal intensity on T1 and T2-weighted sequences. No gadolinium enhancement is visible, in contrast to VMs.

Indication for treatment

Treatment should be considered for lesions located near vital organs and anatomic structures that threaten vital functions, with priority given to respiration, vision, hearing or eating. Early treatment should also be considered for lesions with accompanying complications, such as lymph leakage, bleeding or recurrent infections or cellulitis. Symptomatic lesions, with or without cosmetically severe deformities or functional disability, such as those on the hand, foot, wrist, and ankle should also be considered for early therapy.

Treatment strategies

Pure LMs are only found in infants and newborns. In the majority of patients, additional VMs may exist - mixed malformations.

Therapy includes:

1. Surgical resection; depending on location, partial excision is acceptable, especially as the first stage of a multistage approach.
2. Sclerotherapy with direct puncture, aspiration of fluid and injection of sclerosing agents, like Doxycyclin, Picibanil (OK-432), or ethanolamine oleate, especially in cases of macrocystic LM. Microcystic LMs are not suitable for sclerotherapy [17].
3. Laser treatment.
4. "Off-label" immunmodulating therapy with Rapamycin or Sirolimus.
5. A combination of the techniques is the best solution



Fig. 5. a. Appearance of a high-flow arteriovenous malformation involving the left ear and face as a red, warm pulsating mass. **b.** AVM of the right arm, resulting in soft tissue and bone hypertrophy with typical length discrepancy.

for most patients and should be performed in specialised centers.

Capillary malformations

Clinical presentation

Capillary malformations (CMs), known as “port-wine stain”, typically present as a light-pink to deep-red macule or patch in a geographical or dermatomal pattern and grow proportionally to the patient. The colour deepens to a darker red, and the lesion thickens with time and becomes more nodular. Soft and bony tissue around these malformations may also hypertrophy. Some are multifocal and associated with syndromes linked to known genetic mutations.

Imaging characteristics

CMs are superficial vascular malformations and diagnosis is clinical. No-flow pattern is seen in US examination. The only indication for MRI is to rule out an underlying AVM or associated complex anomalies, such as Sturge-Weber or Klippel-Trenaunay syndromes.

On MRI, CMs may be seen as strongly enhanced thickening of the cutis. However, as they are superficial lesions, they are not always detectable on MRI.

Treatment strategies

Treatment of CMs is directed to the reduction of skin discolouration with serial pulsed dye laser treatment. For extensively thickened lesions and nodules, surgical excision can be performed.

High-flow Malformations

Arteriovenous malformations

Clinical presentation

The vast majority of arteriovenous malformations (AVMs) are sporadically occurring single lesions of unknown aetiology [18]. They are present at birth but may become symptomatic by hormone changes during adolescence and pregnancy or after a trauma [19].

Dermal AVMs may appear as a faint pink patch in neonates but enlarge to form cutaneous lesions. A mature subcutaneous AVM may manifest as a warm, enlarged pulsating mass (**Fig. 5a**). AVMs often cause soft tissue and bony hypertrophy [20], resulting in length and circumference enlargement of the affected limb (**Fig. 5b**). The soft tissue hypertrophy results in asymmetry and, in time, can cause disfigurement. Due to intense

pain, the patients tend to rest the limb, which leads to reduced mobility and additional functional limitations. Clinically, local tissue ischaemia due to arterial steal is relatively common [21, 22].

AVMs tend to progress over time, as collateral arterial flow is recruited into the low-resistance vascular bed. For the same reason, they often recur with a vengeance if incompletely excised or inadequately embolised.

Imaging characteristics

A fast-flow pattern with typical pulsatile arterial pattern, which includes monophasic and triphasic waves, is observed in US. MRI shows arteries and veins with low-signal intensity due to flow void phenomenon of rapid and/or turbulent flow on T1- and especially on T2-weighted spin-echo sequences (**Fig. 6**). Except for the occasional presence of fat, no soft tissue mass is visible. Occasionally, there are numerous small punctuated areas of high-signal intensity caused by haemorrhage and thrombosis. Typical AVMs show gadolinium enhancement only inside the vessels (**Fig. 6**). Gadolinium-enhanced MR angiography is helpful for evaluating feeding arteries and draining veins (**Fig. 7**). The presence of early venous filling is typically seen in AVMs. Using time-resolved MR angiography sequences, it is possible to evaluate the dynamic opacification of AVM with the arterial feeders, nidus and draining veins.

Digital Subtraction Angiography (DSA) is diagnostic and essential for treatment planning in high-flow AVMs [23]. The pathognomonic findings of AVM at DSA are represented by enlarged feeding arteries with an early opacification of the dilated efferent veins. Selective and superselective injections are essential to assess the extent of the AVM and to obtain the exact vascular map, with a focus on feeding arteries and venous drainage, in order to plan the appropriate therapy (**Figs. 7, 8**).

Embolisation

The aim of embolisation is, theoretically, the closure of the nidus. Ideally the closure will include both its in- and outflow zone, but without occluding vessels feeding normal tissue. However, in large peripheral AVMs and multiple nidi, complete cure is the exception. The main therapeutic goal in the endovascular treatment of AVM is embolisation using liquid embolic agents to achieve

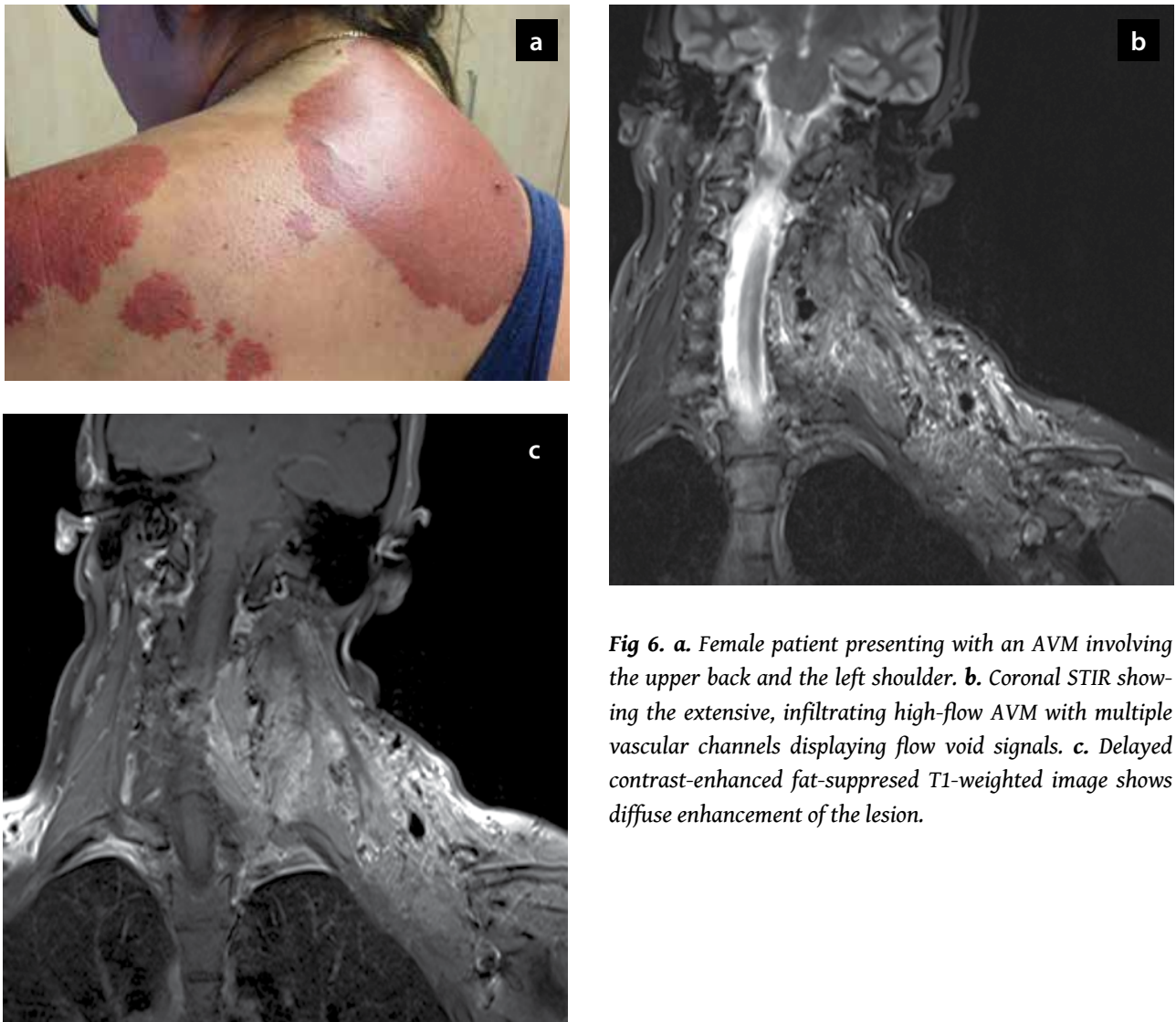


Fig 6. *a.* Female patient presenting with an AVM involving the upper back and the left shoulder. *b.* Coronal STIR showing the extensive, infiltrating high-flow AVM with multiple vascular channels displaying flow void signals. *c.* Delayed contrast-enhanced fat-suppressed T1-weighted image shows diffuse enhancement of the lesion.

a casting of the whole nidus. Most frequently the liquid embolic material used is ONYX, consisting of ethylene vinyl alcohol copolymer dissolved in various concentrations of dimethyl sulfoxide (DMSO) and suspended micronised tantalum powder to provide contrast for fluoroscopy [24]. Depending on the angioarchitecture, a sole transarterial or transvenous approach or the combination of both might be necessary. In most cases the groin is punctured. A transarterial approach can be successful only when reaching, superselectively, the vascular network (nidus) (**Fig. 8**). Proximal occlusion of the feeding arteries is an undesired effect leading to angioneogenesis and revascularisation of the AVM by new, tinier and more tortuous collateralising vessels. In the case of an AVM nidus with a single outflow vein,

retrograde transvenous coil embolisation of that vein directly at the nidus can induce complete retrograde thrombosis and occlusion of the nidus.

Percutaneous embolisation with direct puncture of the nidus can be alternatively used. In percutaneous embolisation (**Fig. 6**) before injecting liquid embolic agents, the correct position of the tip of the needle must be controlled by an angiographic run. The operator must ensure that the tip of the needle is located within the nidus and not in the vein or in the adjacent tissue before deployment of the liquid embolic agent. This technique allows treatment especially in complex AVMs with tiny and tortuous feeding arteries. In our experience, we use this technique often as the primary approach.

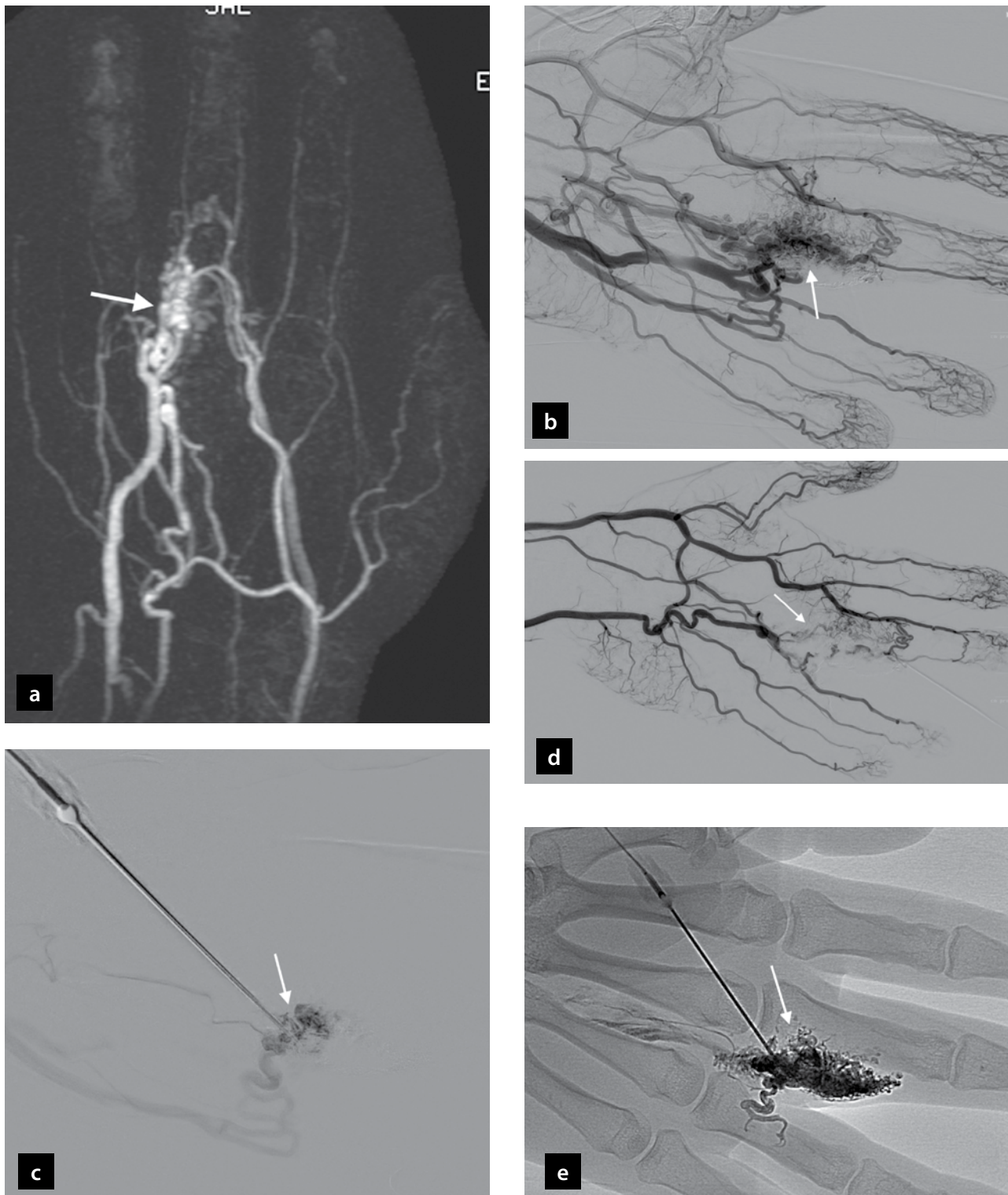


Fig 7. Percutaneous direct puncture of a congenital AVM (arrow) of the right hand of a young female patient (frontal projection). **a.** VIBE image after gadolinium injection showing both digital arteries feeding the AVM (arrow) and the nidus. The dilated draining veins are well demonstrated. **b.** DSA pre-embolisation showing a diffuse AVM (arrow) involving mainly the third finger. **c.** DSA demonstrating the percutaneous direct puncture of the AVM nidus (arrow) in the proximal phalanx of digit III. **d.** DSA post embolisation shows a significant reduction of the nidus (arrow). Note that the preexisting early draining veins are not visible after the embolisation. **e.** Non-subtracted image showing the direct puncture with the ONYX cast in the nidus (arrow) of digit III.

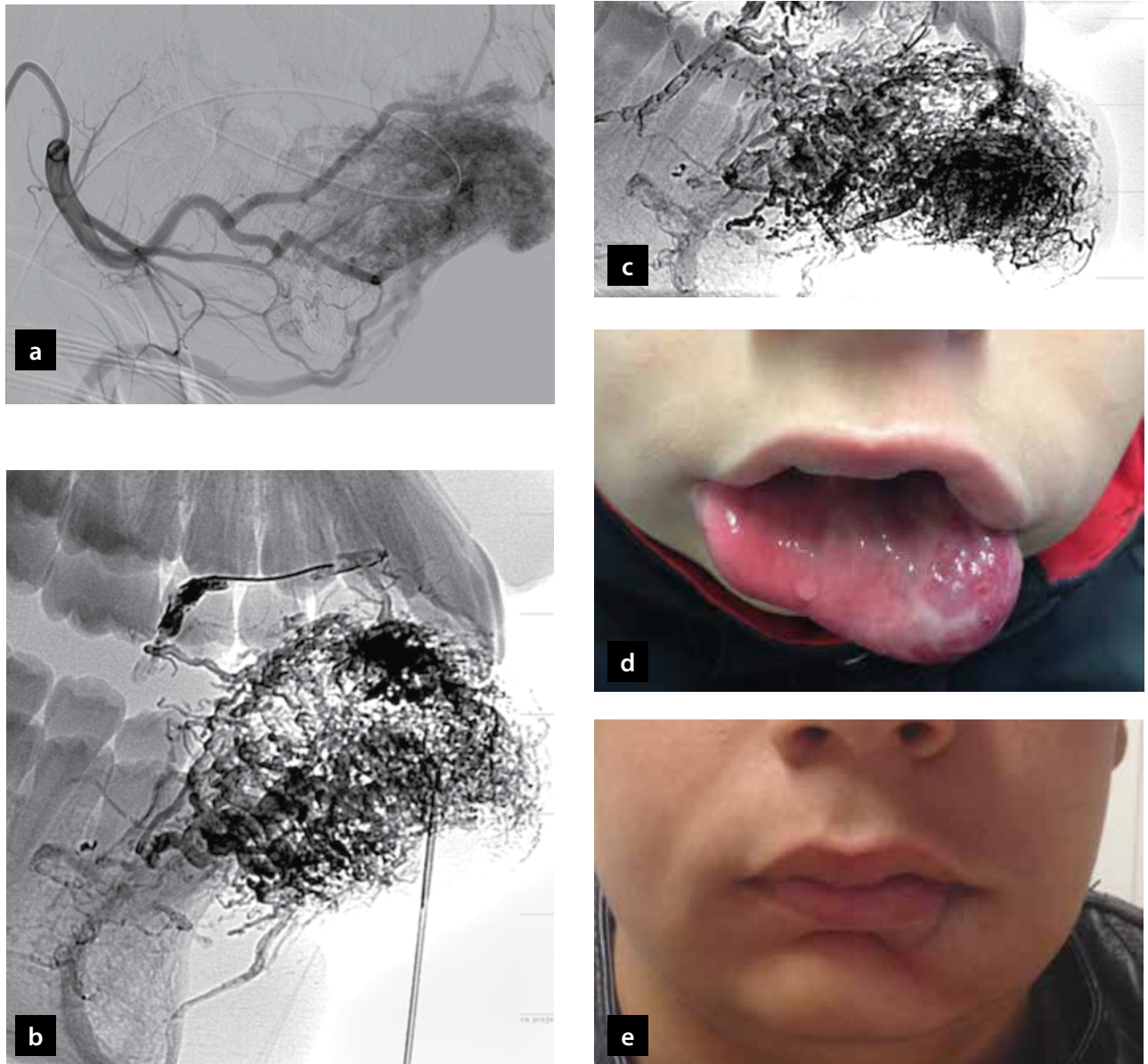


Fig. 8. Embolisation of a diffuse AVM in a young male patient of the lip. **a.** DSA pre-embolisation showing a diffuse nidus with early draining veins. **b.** percutaneous direct puncture of the nidus and embolisation with ONYX. **c.** Non-subtracted image after Embolisation showing the ONYX cast within the nidus. AVM before **(d)** and after **(e)** treatment with embolisation and surgery.

Results

Long-term results in large series are still lacking due to the rarity of the disease. However, patient satisfaction and quality of life are the two most important outcomes. We believe that the first aim should not be to occlude the vessels but rather to improve, or at least stabilise, the physical comfort of the patients. Multiple treatment sessions are nearly always required and a complete “cure” of this disease is only the exception.

Support therapy

Pain control is mandatory for good life quality of the patients. Venous and lymphatic oedema may be significantly reduced by wearing elastic stockings.

Arteriovenous fistulas

Arteriovenous fistulas (AVFs) are often traumatic or iatrogenic acquired lesions and have one or more large AV shunts without a nidus. They are mainly located intracranially and in the neck [25]. They may

become symptomatic with pulsate tinnitus, neurological deficits or bleeding.

The therapy of choice is embolisation with liquid embolisation [ONYX], or surgery, depending on location. This entity is mainly considered a disease of the central nervous system and therefore it will not be further discussed here. Congenital AVFs are rare. Multiple large AVFs are seen in Morbus Rendu-Osler-Weber disease (hereditary haemorrhagic telangiectasia) in the lung and the liver. The major complication is recurrent bleeding. Small lesions throughout the body are treated with laser therapy, while for larger AV shunts in the lung arterial embolisation is indicated [26].

Conclusion

Vascular malformations are complex lesions. They

can be divided based on haemodynamics on “low-flow” and “high flow” malformations. They may be congenital or acquired. Congenital vascular malformations usually become symptomatic during later childhood and adolescence. Acquired vascular malformations consist of post traumatic arteriovenous fistulas. They do not spontaneously regress and they may evolve to disabling, or even life threatening, diseases for the patients. The aim of therapy is to reduce severe symptoms or complications of the lesions. Because of the complexity and rarity of these diseases, they should be treated in specialised centers where a multidisciplinary approach is available. **R**

Conflict of interest

The authors declared no conflicts of interest.

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