Arterio-Venous Malformations (AVMs): imaging findings in a wide spectrum of localizations.

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Learning objectives

We proposed to attend this objectifs:

- **Illustrate the different modalities of clinical presentation and the imaging findings** in patients with Arterio-Venous Malformations (AVMs).
- **Discuss and underline the added value of imaging in diagnosis and in clinical management.**
**Background**

AVMs are rare vascular lesions (ranging from 0.3% to 0.5% of the population) representing errors in vascular development resulting in dysmorphic arterial and venous vessels connected directly to one another without an intervening capillary bed (Fig. 1).

They are present at birth (60%) or become so during the first few weeks of life (30%) with no difference between males and females. Contrary to haemangioma, they never regress and may grow during lifetime.

AVMs may develop during early fetal period, because of the failure of regression of arterio-venous channels in the primitive retiform plexus. This theory explains the predominance of AVMs in the head and neck region, since the early embryo is composed mainly of cephalic structures, having the higher surface area-to volume ratio than other facial structures.

AVMs are frequently sporadic, but they may be associated with underlying disease or systemic anomalies whose molecular genetics have been correlated with AVMs (e.g. hereditary hemorrhagic telangiectasia). Sporadic lesions may include a Transforming Growth Factor b (TGF-b) (it is involved in the induction of apoptotic endothelial cell death) and the tyrosine kinase receptor tunica internal endothelial cell kinase-2 (Tie-2) (it is essential for early vessel development and its increased activity can lead to abnormal growth of the primary vascular plexus) mutation (Fig. 2).

In descending order of frequency they also involve lower and upper limbs, trunk, and viscera (e.g. liver, chest, pelvis) (Fig. 3).

**Histologically**, AVM is composed of multiple dysplastic feeder arteries and arterialized veins creating a vascular nidus without a capillary network. Endothelium cultures show increased growth and reduced apoptosis, suggesting an intrinsic cellular defect. Generally, their histological extension exceeds the visible extension, with microscopic infiltration of the underlying tissue that favours relapse after partial removal (Fig. 4).

**Identification and classification** of vascular anomalies are very difficult and confused; the use of confusing nomenclature continues to persist in the Literature. The accepted classification including AVMs is the International Society for the Study of Vascular Anomalies (ISSVA)/Mulliken's classification modified in 1996. Moreover, vascular malformations can be subdivided on the basis of their vascular components and flow characteristics (slow-flow capillary, venous, or lymphatic channels, fast-flow arterial channels, or a combination of each) and from this point of view, AVMs are considered fast-flow lesions. They are also frequently classified by eponyms when they are components of syndromes (Fig. 5).
The classification is useful to recognize and to study the lesions; in fact AVMs are diagnosed by clinical findings (patient’s medical history and a physical examination) and radiologic features (as a complementary tool specially when there is doubt about the nature of the lesion and as an integral part of treatment).

**Clinical presentation** is extremely variable: asymptomatic lesions, warmer and sometimes pulsatile macules, ulcerated, painful and bleeding lesions, high blood flow lesions; hypertrophy of the bone underlying the lesion is also common. They can be located in critical sites having systemic repercussions on diagnosis and treatment of these lesions. In fact, a proximal arterio-venous malformation with high blood flow may increase cardiac load and lead to congestive heart failure even if there is usually a compensation for years; if the malformation is distal, there is a propensity to lower flow and peripheral ischemia (Fig. 6).

**The natural history** of AVMs can be divided into different stages based on Schobinger staging system (Fig. 7):

1. **Quiescent phase**: asymptomatic lesions or pink-violaceous marks sometimes with a bruit or a thrill if there is a fast-flow component.
2. **Expansive phase**: as in stage I, but clinically pulsatile, with tortuous vessels and tight turns, sometimes invading deep structures.
3. **Destruction phase**: as in stage II with dystrophic skin changes, ulceration, bleeding, and continuous pain.
4. **Decompensate phase**: similar to stage III, with heart failure.

**Stages progression** seems to be favoured by hormonal changes (puberty), pregnancy, traumatic injuries creating a local ischemia, ligation of arterial feeders and partial surgical excision. Local or diffuse soft-tissue deformity, dysmetria, compression, invasion and/or destruction of deep structures, chronic venous deficiency with interstitial edema, cutaneous gangrene can complicate all the lesions (Fig. 8).
Fig. 1: AVMs: definition of pathology.

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Pathologic and genetic features

AVMs are diffuse or localized defects in vessels embryonic development with multifactor genetic component

- chromosomal abnormalities (sporadic mutations)
- involvement of angiogenic factors during embryogenesis (TGF-β, Tie-2)

Fig. 2: AVMs: pathologic and genetic features.

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Fig. 3: AVMs: anatomic localizations.

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Fig. 4: AVMs: pathology.

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**Fig. 5:** AVMs: International Society for the Study of Vascular Anomalies/Mulliken’s classification.

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DIAGNOSIS
clinical characteristics

- Asymptomatic
- Local or diffuse soft-tissue swelling ± visible vessels or ulceration and necrosis
- Palpation: increased warmth, pulse or thrill can be present
- Auscultation: continuous murmur with systolic reinforcement
- Bone and soft tissues hypertrophy because of hypervascularization that sometimes produces severe anatomical deformations

Fig. 6: AVMs: diagnosis - clinical characteristics.

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Fig. 7: AVMs: diagnosis - natural history.

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Complications

- Local or diffuse soft-tissue deformity
- Dysmetria (extremities)
- Dysmetria and deformity (head and neck)
- Compression, invasion and/or destruction of deep structures
- Chronic venous deficiency with interstitial edema
- Skin changes with necrosis, ulceration and gangrene
- Hemorrhage
- Lytic bone lesions
- High-output cardiac failure

**Fig. 8:** AVMs: complications.

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**Imaging findings OR Procedure details**

**Radiologic evaluation is often necessary to confirm the diagnosis, delineate the extent of the lesions, assess the flow characteristics and as a therapeutic option.**

Plain radiography has today a limited value, demonstrating bone involvement (asymmetric hypertrophy or atrophy, osteoporosis, or lytic lesions).

Ultrasonography (US) and colour Doppler evaluations are often performed initially because of their non-invasivity and accessibility and they not require ionizing radiation and a great deal of cooperation of the patient (e.g. paediatric patients). Even if they are operator-dependent techniques, colour Doppler ultrasound is able to confirm vascular nature of the lesion assessing flow characteristics and visualizing multiple internal, well-defined anechoic structures. Colour Doppler analysis can also identify both feeding and draining vascular flow patterns. Spectral waveforms of feeding arteries indicate low peripheral resistance and dilated draining veins show pulsatile flow, suggesting the presence of direct AV communications without an intervening capillary bed. The nidus is characterized by a "mosaic" pattern with a mixture of red and blue colour patterns in the anechoic structures as well as coarse "rumbling" acoustic Doppler representation (Fig. 9, 10).

Computed tomography (CT) is important to visualize vascular components and to define the extent of the AVM. AVMs demonstrate multiple enlarged feeding arteries with rapid contrast shunting into enlarged draining veins without intervening tissue enhancement. Because of radiation, CT ideally should be reserved for particular cases (acute bleeding or adjacent structure compression) (Fig. 11, 12, 13, 14, 15, 16, 25).

Magnetic Resonance (MR) is a non-invasive technique, without ionizing radiation, that provides both anatomic and hemodynamic data. It is excellent for tissue differentiation and this, together with its capacity to acquire images in multiple spatial planes, makes MR the best radiologic technique for demonstrating anatomic relationships of the lesions considering the adjacent structures (muscle and fascial planes), the involvement of bony structures, and providing better detail in confined locations (orbit, lip).

MR shows multiple hypertrophied arteries and dilated veins connected by linear or focal shunting seen as low signal on T1- and T2-weighted spin echo sequences and a characteristic lack of soft tissue component.

On T1- and T2-weighted sequences, the presence of rapid or turbulent flow decreases the intensity of the signal (flow void phenomenon), and when the flow is slow or thrombosis is present, the intensity of the signal increases. In addition, gadolinium enhancement
delineates feeding arteries and draining veins well. MR using phase-contrast (PC) and time of flight (TOF) techniques can successfully identify abnormal arteries and veins. PC-MR is usually sufficient to identify a high flow lesion. Dynamic contrast opacification of the lesion can be performed using time-resolved MR sequences (TRICKS), but spatial resolution tends to be compromised (Fig. 17, 18, 19, 21, 23, 31, 32, 33, 34).

**Angiography** is necessary both to assess the extent of the lesion before therapeutic intervention and to guide intra-arterial embolization; it is specially recommended when MR examination is equivocal or if vascular intervention is considered. It is rarely used for diagnostic purposes alone.

The classic angiographic appearance of AVMs demonstrates multiple hypertrophied feeding arteries rapidly shunting into engorged dilated draining veins via a nidus that is the point at which arterial structures first opacify the venous drainage. No soft tissue enhancement is seen in AVMs, unlike vascular tumours and haemangioma. Direct arterio-venous fistulous components and intralesional aneurysms also may be identified (Fig. 20, 22, 24, 26, 35, 36).

Cho et al. propose an angiographic classification of AVMs based on nidal morphology with implications for therapy and outcomes:

- **Type I**: arterio-venous fistulae
- **Type II**: arteriolo-venous fistulae
- **Type III a**: arteriolo-venulous fistulae with non dilated fistula
- **Type III b**: arteriolo-venulous fistulae with dilated fistula

**AVMs treatment** is complicated, but in all cases the main goal is to obtain complete eradication of the nidus, that causes the high-flow shunting between arterial and venous system. Partial treatment usually results in recurrences that may be more difficult to manage than the initial malformation. Large, diffuse, intracavitary and infiltrating lesions with muscle involvement are inoperable or require extensive, potentially disfiguring, resection or even amputation. Transcatheter and percutaneous nidal embolization often is the first therapeutic option and is an effective approach that can be used as a palliative procedure, giving only a temporary control, or as an adjunct to a surgical resection.

Interruption of the proximal supplying vessels inevitably results in the development of a collateral arterial supply and an inability to access the feeding vessels for endovascular intervention. The use of particulate agents (polyvinyl alcohol particles, PVA), should be reserved for preoperative embolization because recanalization rates are high. A selective or superselective nidal access for embolic agent delivery must be obtained to maximally exposed the nidus to chosen agent effects, minimizing local and systemic complications.
To achieve this goal, sclerosant agents can be delivered transarterially in close proximity to the nidus, using a superselective microcatheter or via a retrograde transvenous approach, with the assistance of a balloon occlusion device; a direct percutaneous puncture into the nidus can also be used.

Moreover, flow reduction techniques, increase concentration, and dwell time allow greater control of distribution of embolic agent within the nidus.

Whatever technique is used, injection of the embolization agent is preceded by contrast injection into the vascular distribution to be embolized to determine the volume and flow rate of the malformation (Fig. 27, 28, 29, 30).
**DIAGNOSIS**

**Imaging modalities**

- Ultrasonography and Color-Doppler (US) evaluation:
  - Heterogeneous lesion with feeding and draining vessels (multiple and well-defined internal anechoic structures)
  - Characteristic high diastolic flow (low peripheral resistance)
  - Arterialization of draining veins (pulsatile flow)
  - Nidus (size, morphology) and multiple sites of arteriovenous shunting

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**Fig. 9:** AVMs: diagnosis - Ultrasonography and Color-Doppler (US) evaluation: a case of posterior and right abdominal wall AVM with multiple and well-defined internal anechoic structures and characteristic high diastolic flow.

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**Fig. 10:** AVMs: diagnosis - Ultrasonography and Color-Doppler (US) evaluation: a case of right forearm's AVM closed to radial artery (25x44x20mm), with typical sonographic appearance.

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**Computed Tomography (CT):**

- AVMs vascular components (multiple enlarged feeding arteries with rapid contrast shunting into enlarged draining vein without tissue enhancement)
- relationship between AVMs and bone structures
- calcifications
- acute bleeding or adjacent structures compression

*Clinical case 3*

![Arterial phase CT scans]

**Fig. 11:** AVMs: diagnosis - Computed Tomography evaluation: arterial phase axial scans show a incidental case of pulmonary AVM (17mm Ø) in the medium lobe (sub-pleural site).

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Fig. 12: AVMs: diagnosis - Computed Tomography evaluation (arterial phase reconstruction: Maximum Intensity Projection-MIP and Volume Rendering-VR): the incidental case of pulmonary AVM (17mm Ø) in the medium lobe; the connection is between the lateral lobe medium artery and the superior pulmonary right vein.

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**Clinical case 4**

**Fig. 13:** AVMs: diagnosis - Computed Tomography evaluation (Maximum Intensity Projection-MIP reconstructions): a case of AVM extending from left iliac fossa to inguinal canal and to scrotal region. Embolic materials of previous treatments are also visible.

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**Fig. 14:** AVMs: diagnosis - Computed Tomography evaluation (axial arterial and venous scans and MPR reconstructions): a case of liver AVM originating from left hepatic artery in a 46 years old male patient in falciform ligament region.

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**Clinical case 6**

![Images of CT scans showing AVMs](image)

**Fig. 15: AVMs: diagnosis - Computed Tomography evaluation (axial arterial scans): a case of ovarian's AVM in a 33 years old patient, extending to cervical uterine region. Vascular connection seems to originate from hypogastric, external iliac and left ovarian arteries and to conclude in hypogastric and ovarian veins. A left cystic ovarian lesion (arrow) and a heterogeneous uterus are also observed.**

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**Fig. 16:** AVMs: diagnosis - Computed Tomography evaluation (Multiplanar-MPR reconstructions): the case of ovarian's AVM in a 33 years old patient, extending to cervical uterine region (54x51x70mm APxLLxCC Ø).

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Magnetic Resonance (MR):

- better assessment of AVMs with respect to adjacent structures (muscle and fascial planes)
- bony structures involvement
- size, morphology and extension of AVMs
- low signal intensity on T2wi because of flow-void phenomenon (rapid and/or turbulent blood flow)
- hemorrhage or thrombosis can be present as small punctuate areas of high signal intensity

Fig. 17: AVMs: diagnosis - Magnetic Resonance evaluation.

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**Fig. 18:** AVMs: diagnosis - Magnetic Resonance evaluation: MR exam confirms the presence of ovarian's AVM extending to cervical uterine region with vascular connection originating from hypogastric, external iliac and left ovarian arteries and concluding in hypogastric and ovarian veins.

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**Clinical case 7**

**Fig. 19:** AVMs: diagnosis - Magnetic Resonance evaluation: superior lip's AVM in a 37 years old female patient (11mm Ø) connected to orbicular muscle. The lesion shows early uptake of contrast medium (high flow lesion probably supplied by superior lip vessels).

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**Angiography:**
- better information on structure, number and size of feeder arteries, nidus and draining veins
- hemodynamic study (artero-venous shunts and connections with other vascular districts)
- possibility to treat (selective and superselective injection are essential to determine the extent of the lesion and to precise vascular anatomy of the feeding vessels)

*Fig. 20:* AVMs: diagnosis - Angiography: a diagram of the angiographic classification of AVMs.

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**Fig. 21**: AVMs: diagnosis - Magnetic Resonance (MR) evaluation: a case of posterior and right abdominal wall AVM in a 37 years old male patient with multiple vascular feeders originating from lumbar vessels.

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Fig. 22: AVMs: diagnosis - Angiographic evaluation and treatment: the case of posterior and right abdominal wall AVM in a 37 years old male patient with multiple vascular feeders originating from lumbar vessels treated with coils super-selective embolization.

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**Clinical case 9**

**Fig. 23:** AVMs: diagnosis - Magnetic Resonance (MR) evaluation: a case of left gluteal AVM in a 38 years old female patient with multiple vascular feeders originating from hypogastric artery and draining into iliac veins.

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Fig. 24: AVMs: diagnosis - Angiographic evaluation: the case of left gluteal AVM in a 38 years old female patient with multiple vascular feeders originating from hypogastric artery and draining into iliac veins.

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Clinical case 10

**Fig. 25:** AVMs: diagnosis - Computed Tomography evaluation (Maximum Intensity Projection-MIP reconstructions): a case of AVM in a 59 years old male patient originating from superior mesenteric artery. After medium contrast administration we observe a early opacification of superior mesenteric portal and paraumbilical veins.

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**Fig. 26:** AVMs: diagnosis - Angiographic evaluation: the case of AVM in a 59 years old male patient originating from superior mesenteric artery. After medium contrast administration we observe a early opacification of superior mesenteric portal and paraumbilical veins.

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Fig. 27: AVMs: treatment.

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**Treatment**

- **Surgical ligation** (small and medium size AVMs)

- **Embolization** and surgical ligation
  
  - Intraoperative bleeding

- **Embolization**: to achieve a temporary control

*Fig. 28: AVMs: treatment options.*

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Treatment
Catheterism and Embolization technique

- Nidal access for embolic agent
  1 Arterious catheterism
  2 Direct percutaneous puncture
  3 Venous catheterism

- Preliminary selective angiographic study is essential:
  - To identify all the feeder arteries
  - To evaluate AVMs high/low flow (choice of embolic agents)
  - To select the target to embolize

Fig. 29: AVMs: treatment: catheterism and embolization technique.

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Coils - Vascular Plug
- +++ high flow AVMs

Polyviyl Alcool Foam Powder (150-500 μm)
- smallest size in order to reach smaller vessels
- particular attention in the case of direct communication AV

Cyanoacrylates
- < risk of recurrence by accumulating in the nidus
- < flow and > concentration to control the distribution of embolic agent within the nidus

High rates of recurrence but temporary control of the lesion

Fig. 30: AVMs treatment: materials.
**Clinical case 11**

Fig. 31: AVMs: diagnosis - Magnetic Resonance (MR) evaluation: a case of right thigh AVM in a 12 years old female patient with multiple vascular feeders originating from deep femoral artery.

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Fig. 32: AVMs: diagnosis - Magnetic Resonance (MR) evaluation: a case of right thigh AVM (9x11x25mm Ø) in a 12 years old female patient with multiple vascular feeders originating from deep femoral artery.

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Fig. 33: AVMs: diagnosis - Magnetic Resonance (MR) evaluation: a case of right thigh AVM (9x11x25mm Ø) in a 12 years old female patient with multiple vascular feeders originating from deep femoral artery.

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**Fig. 34:** AVMs: diagnosis - Magnetic Resonance (MR) evaluation: a case of right thigh AVM in a 12 years old female patient with multiple vascular feeders originating from deep femoral artery. A early opacification of the draining venous vessels is present.

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Fig. 35: AVMs: diagnosis - Angiographic evaluation and treatment: diagnostic and therapeutic procedure in right thigh AVM 12 (coils super-selective embolization).

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Fig. 36: AVMs: diagnosis - Angiographic evaluation and treatment: diagnostic procedure in right thigh AVM after treatment, shows a partial reduction of the lesion.

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Vascular malformations are highly complex lesions that require a highly specialized multidisciplinary approach.

Early diagnosis and appropriate treatment are crucial because these lesions can cause many serious problems such as congestive heart failure and acute bleeding.

Radiologic evaluation (Doppler US, CT, MR imaging) is necessary to confirm the diagnosis, delineate the lesion extent and assess the flow characteristics. Interventional radiology is useful to confirm the diagnosis and to manage the lesion as therapy of choice or as an adjunct to surgery.
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