# How to manage pelvic arterio-venous malformation

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Learning objectives

1. To learn clinical features of pelvic AVMs
2. To learn imaging features of pelvic AVMs
3. To learn treating strategies
4. To learn complications and outcomes of each treatment.

Background

Congenital pelvic arteriovenous malformations (AVMs) are rare diseases that are seen mainly in women aged 30-50 years. They progress slowly over time and can remain asymptomatic for years. However, the symptoms can be incapacitating or even life-threatening. They are treated with surgical resection, embolization, or a combination of both techniques, although the recurrence rate is high. The etiology and clinical behavior of pelvic AVMs are quite variable, and therapy should be individualized based on the clinical and pathological conditions of each case.

In this study, we describe the clinical features of pelvic AVMs and introduce a treatment strategy based on the angiographic patterns of the disease.

Imaging findings OR Procedure details

1. Clinical features of pelvic AVMs

Congenital pelvic AVMs result from mal-development of arterial, capillary, venous, or lymphatic structures. Multiple macro-communications and micro-communications between the arterial and venous systems with resultant shunting of blood to the low-resistance veins produce massive venous and tissue engorgement. The cause of presenting symptom is related to venous hypertension.

Common symptoms

I Abdominal or pelvic discomfort and pain: most common (44%)
I Rectal pain and tenesmus
I Genitor-urinary symptoms: hematuria, hydronephrosis, unusual vaginal bleeding
**Rare symptoms**

- High-output cardiac failure: 20%
- Sciatica

**Physical examination**

- A pulsatile mass palpated during abdominal or rectal examination: most common (78%)
- Continuous bruit and thrill auscultated in the groin, radiating to the umbilicus.
- Edema of the lower extremities

**2. Imaging features of pelvic AVMs**

Pelvic arteriovenous malformations may be supplied by any artery in the pelvis but the primary feeders, markedly dilated and tortuous, seem to be gonadal arteries, branches of the internal iliac arteries, hemorrhoidal branches of the inferior mesenteric artery, and the middle sacral artery. Most often a hypertrophied internal iliac artery supplies a dense nidus of the AVM, which then shunts almost like a fistula into massively dilated veins.

**Computed tomography (CT) with bolus contrast enhancement:**

- Delineates the location and extent of the disease,
- Delineates the involvement of adjacent structures, and
- Can identify the feeder and drainage vessels from close observation of thin slices and three-dimensional images.

**Magnetic resonance imaging (MRI):**

- Delineates the location and extent of the disease,
- Delineates the involvement of adjacent structures,
- Can distinguish slow-flow venous malformations from high-flow AVMs and arteriovenous fistulas (AVFs), and
- With intravenous minus intra-arterial subtraction MR angiography technique, can delineate additional feeding branches not clearly identifiable at nonselective pelvic angiography (Ugur B. et al. J Vasc Interv Radiol 2007; 18:920-923)

**Echo-Doppler examination:**

- Confirms the presence of an AVM,
Delineates flow distribution, and

Is easy to perform and useful for monitoring

**Angiography:**

Delineates both the size of the AVM and the extent of vessel involvement, and

Delineates tortuous, hypertrophic feeding arteries and dilated drainage veins with a tangle of poorly defined, serpiginous arteries interposed, which are characteristic features of pelvic AVM.

### 3. Treating strategies

**Classification of pelvic AVMs for therapeutic purposes**

Olcott et al. classified AVMs for therapeutic purposes, and we generally agree with their proposed classification. (Surgery 1976; 79: 3-12)

**Type 1:** The nidus is confined to a small area and can be removed safely; surgical resection, not ligation, is used.

**Type 2:** The nidus is more extensive, making its removal difficult but not impossible; embolization is often performed before surgery.

**Type 3:** The nidus is extensive, making its removal impossible. Repeated embolization is required because there are repetitive cycles of symptomatic remission and relapse with this type.

Of all symptomatic pelvic AVMs, only a few are type 1. Furthermore, we believe that embolization may be tried initially because of its lower mortality compared with surgery. Most symptomatic patients are usually type 2 or 3, and require embolization. Advanced treatment techniques and new devices can achieve complete remission in some of these cases.

Houdart et al. classified high-flow intracranial AVMs based on their angiographic appearance. This classification also be applied to pelvic AVMs. (Neuroradiology 1993; 35:381-385)

**Type 1:** Arteriovenous AVMs, in which no more than three separate arteries supply a single initial venous component.
Fig.

**Type 2:** Arteriolo-venous AVMs, in which multiple arteries shunt into a single, central, dilated venous component.

Fig.

**Type 3:** Arteriolo-venulous AVMs, in which there are multiple shunts between arterioles and venules. In this type, the first identifiable normal venulous component is separate from the shunts.
**Type 4, which we have added:** In most large pelvic AVMs, complex combinations of these three types are present.

For all of these types, embolization must be performed as near as possible to, or across, the arteriovenous communications (i.e., the niduses) themselves. Unduly proximal embolization of feeding arteries is to be avoided, because, as in the case of surgical ligation, recurrence is invariable and subsequent access to the lesion for more definitive treatment will be severely compromised.

**Recommendation of access route in each type**

**Type 1**

I An arterial approach is recommended.

I There are few targeted feeders, and it is comparatively easy to approach the arteriovenous communication in this type.

I Adequately sized coils, microparticles, or some sclerosing agents such as N-butyl-2-cyanoacrylate (NBCA) are used.
Fig.

Type 2

An initial venous approach is recommended, because it is difficult to catheterize and embolize the numerous feeders from the arterial side. As the efferent vein is dilated in this type, the catheter can approach the arteriovenous communication.

When occlusion of the efferent veins is difficult, repeated embolization from the arterial side is subsequently performed.

Liquid sclerosing agents such as ethanolamine oleate (EO) or absolute ethanol are used from the venous side, whereas NBCA is recommended for use from the arterial side.

Fig.

Type 3
Repeated embolization from the arterial side is indicated.

When AVMs becomes localized after repeated embolization and the efferent veins can be occluded, venous approach is indicated.

As the efferent veins are usually dilated, a microcatheter can easily approach the nidus.

NBCA is recommended for injection from the feeding arteries; EO is preferred for the venous approach.

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**Fig.**

**Type 4**

Initially, the transarterial approach is used, and the AVF components are embolized with coils, NBCA, or microparticles.

Repeated embolization with NBCA from arterial side should be added.

Finally, embolization is performed from venous side with a liquid sclerosing agent such as EO.

**Embolic materials**

**Coils/microballoons**

These materials are not used initially except AVF components, because they often embolize the proximal region unduly, which leads to recanalization. Large, complex AVMs often contain large arteriovenous communications such as AVFs. The flow velocities through these components are very high, and they must be embolized carefully with adequately sized coils or microballoons. Other embolic materials risk being embolized to the lungs in such cases.
Microparticles

Polyvinyl alcohol foam particles (Ivaron) have been used for years, although several authors have suggested that the particulate embolization of pelvic AVMs is a palliative procedure, because of the frequent recurrence. The large particles embolize proximally, small ones pass through arterivenous communication.

Fig.

Absolute ethanol

Absolute alcohol is a dehydrating agent and can strongly thrombose the vessels it contacts. Arterial flow control is necessary to expose the vessel wall to this agent.

Careful management is needed because unexpectedly wide spread of the agent causes ischemic complications, tissue degeneration, or neuralgia.
N-butyl-2-cyanoacrylate (NBCA)

A mixture of cyanoacrylate and lipiodol is often used initially to embolize pelvic AVMs. This agent has the potential to embolize the nidus with afferent arteries. The complete embolization of a pelvic AVM may be achieved, especially when the feeding arteries are limited to a few dilated vessels.

Fig.

The ratio of NBCA to lipiodol determines the polymerization time, which enables adequate sclerosis at or around the nidus. In the case of high-flow AVMs, the ratio of NBCA to lipiodol may be 2:3 or 1:1 in order to shorten polymerization time.

In cases of slow-flowing AVMs or a long distance between catheter and nidus, the ratio may be reduced to 1:2 to 1:3 in order to slow polymerization.
A mismatch between the blend ratio and the flow velocity of AVMs leads to unexpected proximal embolization, nidul hypertension, or pulmonary embolization.

**Fig.**

*Ethanolamine oleate (EO)*

Benzyl alcohol containing EO affects the endothelial cells of the vessels, resulting in the activation of coagulation factors and platelet aggregation. Therefore, to embolize AVMs, the nidus should be exposed to EO for some time. When EO combines with albumin, the platelets-agglutinating action of EO rapidly dissipates. Therefore, in the event that EO flows into a distant vessel, thrombi are not produced. We believe that EO injection from a microcatheter at or near the nidus, with occlusion of the efferent vein, is the best way to treat pelvic AVMs.
4. Complications and outcomes

**Complications**

**Common**

- **Pain:** Ranging from vague discomfort to severe pain requiring narcotics.

- **Low grade fever:** Well correlated with the extent of the embolized territory

- **Pulmonary embolization:** After embolization of high-flow intracranial AVMs with NBCA, a plain chest x-ray showed whiteness due to the sclerosing material in 24% of cases, indicating pulmonary embolization. Symptomatic embolization may occur in 1 to 2%.
Rare

- Venous thrombosis: May occur in a few cases, and some of them exhibit the symptom of stasis or venous hypertension.

- Ischemia: Unexpected embolization of some important artery may cause ischemia. Repeated embolization may elevate the incidence of ischemia.

- Difficulty in bowel movement: Due to blood stasis or venous hypertension

- Sciatic nerve palsy: Due to ischemia or infiltration of absolute ethanol

Clinical outcomes

- The short-term outcomes are good, but long-term outcomes are poor because of the high incidence of recurrence (84%).

- Complete embolization of the arteriovenous communication is the key to preventing recurrence.

- Many authors agree that the treatment of large pelvic AVM is palliative, although recently developed techniques enable us to completely cure some of these cases.

5. Our experience

We have treated five patients with pelvic AVMs in 15 years. With recurrences, 11 procedures were needed. In the initial cases, we used microcoils and absolute ethanol from the arterial side; more recently, we preferred to use NBCA.

One patient underwent complete embolization with NBCA in one session and has been free of relapse for 9 years.

In our most recently treated case, who had been treated three times previously, we successfully embolized every nidus with EO injected from the venous side.

The three other three patients have undergone years of cure and recurrence; one died of thyroid cancer during follow-up.

6. Case reports

Case 1

A 63-year-old female suffered from left lower abdominal pain and high-output cardiac failure.

A) CT revealed a large pelvic AVM in the left pelvis.
B) Digital subtraction angiography (DSA) demonstrated multiple niduses, and the AVF component in the region was supplied by the internal iliac artery bilaterally. After embolization with microcoils and absolute ethmol, some nidal flow still remained (large arrow).
C) Her symptoms recurred within 4 months. DSA showed recanalization of multiple niduses and engorged drainage vein, although AVF component was still occluded (arrowhead).
Fig.

D) Multiple feeders were embolized with a NBCA:lipiodol at 2:3, although a few afferent vessels remained (large arrow), followed by the residual dilated efferent vein.
**Fig.**

E) A plain chest plain x-ray revealed whiteness in the right lung (small arrows), which proved to be NBCA that had embolized to the lung. Note that the cardio-thoracic ratio was reduced after therapy.
The patient had remained asymptomatic 1 year after last embolization, when she died of thyroid cancer.

**Case 2**

A 68-year-old male suffered from intermittent lower gastrointestinal bleeding

A) CT revealed a large localized pelvic AVM in the left pelvis.
B) DSA showed three or four dilated afferent arteries, followed by multiple niduses (large arrow). Two dilated drainage vein are also seen. After transarterial embolization (TAE) with NBCA, the afferent arteries and nidus became column-shaped. The ratio of NBCA to lipiodol was 2:3.
Fig.

C) After 6 month, no residual AVM was seen on DSA.
Fig.

There has been no relapse after 9 years.

Case 3

A 70-year-old male suffered from recurrent lower abdominal pain and intermittent gastrointestinal bleeding.

A) TAE has been performed three times before this visit. Multiple branches of the internal iliac arteries bilaterally and inferior mesenteric artery had been embolized with NBCA, absolute ethanol and microcoils.
Fig.

B) CT revealed a large efferent vein and multiple nidus from the peripherals of multiple small arteries. There were multiple niduses efferent to a large vein, which branched to some dilated branches. Transvenous embolization was planned, as the transarterial approach is difficult and risky in cases after multiple embolizations.
Fig.

C) The dilated gluteal vein (red arrow) was punctured under ultrasonography guidance, because there was severe stenosis (yellow arrow) in the right internal iliac vein, which made it difficult to insert a balloon catheter into the efferent vein from the left common femoral vein.
Fig.

D) A 7-Fr balloon catheter was inserted and used to occlude the efferent vein. Then, a microcatheter was inserted to the region near the nidus.
E) After occlusion of the efferent vein, 10 ml of EO were injected in pulses from the microcatheter to the exposed nidus. The nidus was visualized clearly. The balloon was deflated after 1 h, and the thrombus did not move.
Fig.

F) Three months after the therapy, no recurrence was seen on CT. The drainage veins were also thrombosed.
After therapy, the patient suffered from transient difficulty with his bowel movements, but this improved slowly over a few months. There was transient elevation of prostate-specific antigen (PSA) and MRI revealed edema of the anal sphincter, which also improved slowly.

**Conclusion**

The choice of therapy for pelvic AVM should be based on the specific types. A localized pelvic AVM may be treated with surgery or by embolotherapy using either a transareterial or transvenous approach. A large AVM should be treated with repeated transarterial embolotherapy. Finally, some recurrent pelvic AVMs can be treated by using transevenous embolotherapy.

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**References**


