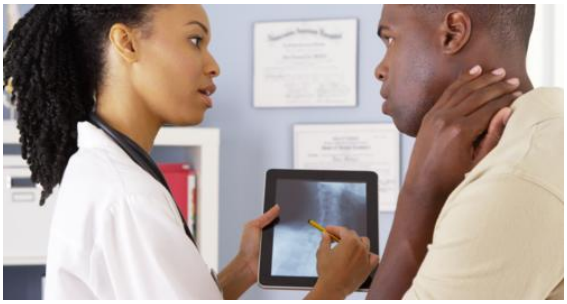
Vascular Malformations



Vascular malformations are non-cancerous growths made up of a tangle of one or more types of blood vessels and can be thought of as a kind of birthmark. Most vascular malformations are present at birth (congenital) and occur by chance (sporadic) similar to a birthmark. Although, there are a few types that can be inherited as a family trait.

Some vascular malformations may be very noticeable, slightly noticeable or not noticeable at birth. Vascular malformations will grow with time. Some vascular malformations can become prominent suddenly due to illness or some may become more prominent gradually during periods of growth such as adolescence or pregnancy.

Vascular malformations can be small, focused in one area of the body, or they can be large, involving several areas of the body. Vascular malformations may range from asymptomatic to seriously symptomatic. Serious symptoms include difficulty walking or moving, pain, swelling, blood clots, muscle weakness, and, if it is located in the head and face, problems with seeing, breathing, or swallowing.

Vascular malformations that are symptomatic may require treatment. Both diagnosis and treatment will depend on your specific type of vascular malformation. There are many different types of vascular malformations that are named based on the vessels that are involved. The four main vessels in our bodies include: lymphatics, veins, arteries, and capillaries.

The three most common vascular malformations (VMs) include lymphatic, venous and arteriovenous malformations.

Lymphatic Malformations (LMs)

What is a lymphatic malformation?

The lymphatic system (http://www.radiologyinfo.org) includes a network of small tube-like vessels that run throughout the body. These vessels collect and carry lymph (http://www.radiologyinfo.org) fluid from tissues and organs, through the lymph nodes and into the blood circulation. This network, called the lymphatic system, is part of the body's immune system.

A lymphatic malformation (LM) (http://www.radiologyinfo.org) happens when a misshapen lymph vessel traps lymph fluid. The trapped fluid forms a cyst, which may grow either gradually or suddenly. Blood from nearby vessels may also enter this cyst, causing the area around the LM to swell. LMs can sometimes get infected and cause swelling. An enlarged LM may result in pressure on nearby tissues and organs. If an LM becomes enlarged or painful, you should see your physician.

LMs can appear anywhere in the body. They are most common on the neck, face, armpit, or buttocks. They may also appear inside the body, in organs or bones.

LMs are congenital, which means the malformation is present at the time of birth. A LM may be found in a fetus during pregnancy. Others may be found later in childhood, when it appears as a fluid-filled lump.

There are three LM types:

- macrocystic

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- microcystic
- mixed (macro and microcystic)

Macrocystic LMs may include one or more cysts that are one centimeter or larger. They are usually soft to the touch. Microcystic LMs are a group of smaller cysts that may feel solid. These may appear anywhere in the body. Mixed LMs may contain macrocysts and microcysts.

LMs are not hereditary. They are not associated with any other medical conditions. However, the condition is more common in children with:

- Down syndrome
- Turner syndrome
- Overgrowth syndromes (genetic disorders that cause an unusual increase in the size of the body or a body part)
- Noonan syndrome.

Symptoms

If you have an LM, your symptoms will depend on the size of the malformation and where it is located. Symptoms may include:

- A soft, smooth lump on the skin that may or may not have discoloration. It may be found anywhere on the body including the neck, head, mouth, tongue, eye, chest, abdomen, arms, legs, scrotum, or penis.
- A lump or mass that gets larger quickly.
- A lump that shows signs of infection, including redness, warmth, pain, swelling and drainage (rarely).
- Chronic small bumps, blisters, or bloody crusts on the surface of the skin that may rupture and ooze blood or clear lymph fluid.
- A LM in the head, neck or tongue region may make it hard to breathe or swallow.
- LMs in an arm or leg may cause swelling and pain throughout the entire limb.

How is a lymphatic malformation diagnosed and evaluated?

Before a baby is born, a LM may be found during an obstetrical ultrasound (US) (https://www.radiologyinfo.org/en/info/obstetricus) or Fetal MRI (https://www.radiologyinfo.org/en/info/fetal-mri) , which provide pictures of a fetus within the uterus.

A LM on the skin may be found during a physical examination. Imaging is needed to confirm the malformation. Imaging is also used to diagnose a LM inside the body.

You may have one or both of these imaging tests:

- Ultrasound: (https://www.radiologyinfo.org/en/info/genus) Ultrasound imaging uses sound waves to produce pictures of the inside of the body.
  For more information about ultrasound performed on children, visit the Pediatric Abdominal Ultrasound (https://www.radiologyinfo.org/en/info/abdomus-pdi) page.
- Body magnetic resonance imaging: (https://www.radiologyinfo.org/en/info/bodymr) MRI uses a powerful magnetic field, radiofrequency pulses and a computer to produce detailed pictures of internal body structures. MRI does not use radiation (x-rays).
  For more information about magnetic resonance imaging performed on children, visit the Pediatric MRI (https://www.radiologyinfo.org/en/info/pediatric-mri) page.

How is a lymphatic malformation treated?

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Not all LMs require treatment. If the LM is not causing problems, you and your doctor may decide to watch it over time. Treating an LM will depend on:

- where it is located in the body.
- what symptoms you are experiencing.
- whether it is macrocystic, microcystic, or mixed.
- whether nearby tissues, blood vessels and/or organs are affected.
- the patient's age (for children), medical history and overall health.

Treatment options include:

- Antibiotic medications to treat an infected LM.
- Compression therapy (http://www.radiologyinfo.org) , or wearing a tight-fitting garment on the affected area of the body. The garment helps prevent the malformation from getting larger and reduces swelling in an arm or leg.
- Draining the cysts. For this procedure, a drainage tube is placed through a small skin opening into the cyst(s) using image guidance.
- *Sclerotherapy (https://www.radiologyinfo.org/en/info/sclerotherapy)* . Your doctor will inject a solution (sclerosant) directly into the malformed lymph vessels. This causes them to shrink and collapse. The doctor begins by draining the cyst(s) using image guidance and then injecting the sclerosant. Sclerotherapy is most often used for macrocystic LMs. Some microcystic LMs also respond to sclerotherapy. Several sclerotherapy sessions may be required.
- Surgery. Your doctor may remove some LMs using surgery. LMs that affect too large an area, such as an entire arm, may not be suitable for surgery.
- Laser therapy. Doctors use a strong beam of light to destroy the LM. Laser therapy is used for LMs of the skin or mouth. Several treatments spaced over several months may be needed. This therapy may be used in combination with other treatments.
- Medical therapy. For extensive or complicated LMs, your doctor may prescribe skin ointments, oral medication, or intravenous medication. Your doctor may also use medication to shrink an LM or to reduce the risk of an LM coming back after surgery or sclerotherapy.

Doctors are investigating other ways to treat LMs, including the use of cold (*cryoablation (https://www.radiologyinfo.org/en/info/thermal-ablation-therapy)*) or heat (radiofrequency or microwave ablation).

Venous Malformations

What are venous malformations (VMs)?

Venous malformations (VMs) (http://www.radiologyinfo.org) are the most common type of vascular malformations. The malformation happens when a tangle of veins grow abnormally in a specific area. The malformed veins can stretch and grow larger over time.

VMs may occur anywhere, from deep inside the body to closer to the skin. The condition may occur in one area or several areas of the body. If the malformation is just under the skin, it may be bluish in color or look like a bruise. If you press on this type of VM, you may feel round, hard bumps. These are calcified blood clots about the size of a pearl called phleboliths. When you press down on the VM, blood empties out of the misshapen veins and returns when you stop applying pressure.

Blood tends to flow through a VM slowly, which may cause swelling and clotting.

VMs may start small early in a child's life and grow when the patient goes through hormonal changes such as puberty or pregnancy.

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Symptoms of a VM depend on its location and include:

- pain
- swelling
- bleeding
- open sores (ulcers) on the skin
- muscle cramping
- joint pain when walking or a feeling of heaviness (if located in the leg)
- difficulty breathing or speaking (if located in the mouth)
- blood in the stool or urine (if located in the intestines or bladder).

How are VMs diagnosed and evaluated?

Your doctor may find a VM on the skin during a physical exam. Your doctor will use imaging tests to confirm the malformation, such as:

Ultrasound (https://www.radiologyinfo.org/en/info/genus) : Ultrasound imaging uses sound waves to produce pictures of the inside of the body.

Ultrasound is easier for children because it doesn't require a child to lie still. It can also be performed without sedation. *For more information about ultrasound performed on children, visit the Pediatric Abdominal Ultrasound (https://www.radiologyinfo.org/en/info/abdomus-pdi) page.*

Magnetic resonance imaging (MRI): (http://www.radiologyinfo.org) MRI does not use radiation (x-rays). Instead, it uses a powerful magnetic field, radiofrequency pulses, and a computer to locate and image malformations. It also shows other important structures, such as nearby nerves, muscles, and arteries that treatment may affect. MR angiography (MRA) (https://www.radiologyinfo.org/en/info/angiomr) may be part of this exam. These imaging tests can show whether there are arteries connecting to the VM or if veins are draining blood from the VM.

For more information about magnetic resonance imaging performed on children, visit the Pediatric MRI (https://www.radiologyinfo.org/en/info/pediatric-mri) page.

How are VMs treated?

A VM that is not causing symptoms or problems does not have to be treated. If a VM is not causing problems, you and your doctor may decide to watch it over time. Most VMs do not require immediate treatment but starting treatment will depend on:

- where it is located in the body.
- what symptoms you are experiencing.
- whether nearby tissues, blood vessels and/or organs are affected.

Treatment options include:

- Compression therapy (http://www.radiologyinfo.org) , or wearing a tight-fitting garment on the affected area of the body. The garment helps prevent the malformation from getting larger and reduces swelling in an arm or leg.
- Medications to treat symptoms and complications.
- *Sclerotherapy (https://www.radiologyinfo.org/en/info/sclerotherapy)* . A doctor injects a solution directly into the malformed blood vessel(s). This causes them to shrink and collapse. You may need several sclerotherapy sessions. Your doctor may surgically remove a VM following sclerotherapy.
- *Embolization (https://www.radiologyinfo.org/en/info/dc-embol) :* using image guidance, the doctor inserts and maneuvers a

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catheter through the body to the malformation. The doctor injects a liquid adhesive through the catheter to seal the vessel. They may also embolize the malformation and surgically remove it.

- Surgery. Your doctor may remove a small VM in a single location using surgery. If the VM is large or near important organs and structures in the body, only part of the VM may be removed.
- Doctors are investigating other ways to treat VMs, including the use of cold (*cryoablation (https://www.radiologyinfo.org/en/info/thermal-ablation-therapy)*) or heat (radiofrequency and microwave ablation).

Arteriovenous Malformations

What are arteriovenous malformations (AVMs)?

In an AVM, blood flows directly from an artery into a vein instead of passing through capillaries. AVMs are often found in the brain, neck, and spine. Peripheral AVMs are found in the arms, legs, and internal organs, including the kidneys, intestines, and lungs.

Most AVMS are congenital or present at birth.

Some AVMs cause no problems. Others may burst and bleed. Most episodes of bleeding are not severe enough to cause permanent damage. However, significant bleeding can occur. A bleeding AVM in the brain may cause a stroke or brain damage. Peripheral AVMs reduce the blood supply to nearby tissue. Over time, this can damage the tissue and cause pain and ulcers (open sores), on the skin. They may also force the heart to work harder to circulate blood.

Vein of Galen Malformations

Vein of Galen malformations (VOGMs) are a type of AVM that may be identified while your baby is still in the womb or immediately after birth.

Symptoms

Children who are born with AVMs may not have symptoms for many years. Symptoms may occur between the ages of 10 and 40. Many AVMs, especially in the head, are not recognized until adulthood.

A child with an AVM may have these symptoms:

- a pink, red, or purple birthmark
- pain
- swelling
- bleeding, which may be difficult to stop
- warmer skin over the AVM
- a pulse that's felt around the AVM.

Symptoms of head, neck, and spine AVMs include:

- headaches
- neck pain
- weakness
- seizures
- an unusual sound, such as humming, pulsations, or swishing, in one ear
- double vision or other visual disturbances

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- increased pressure in the eye (glaucoma)
- eye swelling, decreased vision, redness and congestion of the eye
- dizziness
- stroke
- seizures
- problems with speech, vision, or movement.
- prominent blood vessels on the scalp and above the ear.

Symptoms of peripheral AVMs include:

- shortness of breath when active
- coughing up blood (if AVMs are in the lungs)
- bleeding
- abdominal pain
- black stools (if AVMs are in the digestive system)
- *anemia (https://www.radiologyinfo.org/en/info/anemia)*
- swelling
- lumps
- open sores (ulcers) on the skin
- pain.

How are AVMs diagnosed and evaluated?

Your doctor may find an AVM on the skin during a physical exam. AVMs are sometimes found during treatment for an unrelated disorder. Your doctor will use imaging tests to confirm the malformation, such as:

Ultrasound (https://www.radiologyinfo.org/en/info/genus) : Ultrasound imaging uses sound waves to produce pictures of the inside of the body.

Ultrasound is easier for children because it doesn't require a child to lie still. It can also be performed without sedation. *For more information about ultrasound performed on children, visit the Pediatric Abdominal Ultrasound (https://www.radiologyinfo.org/en/info/abdomus-pdi) page.*

Body (https://www.radiologyinfo.org/en/info/bodymr) and head (https://www.radiologyinfo.org/en/info/mri-brain) magnetic resonance imaging (MRI): MRI does not use radiation (x-rays). Instead, it uses a powerful magnetic field, radiofrequency pulses, and a computer to locate and image malformations. It also shows other important structures, such as nearby nerves, muscles, and arteries that treatment may affect. MR angiography (MRA) (https://www.radiologyinfo.org/en/info/angiomr) or MR perfusion (MRP) may be part of this exam. These imaging tests can show the arteries connecting to the AVM or the veins that are draining blood from the AVM.

For more information about magnetic resonance imaging performed on children, visit the Pediatric MRI (https://www.radiologyinfo.org/en/info/pediatric-mri) page.

Head computed tomography (CT) (https://www.radiologyinfo.org/en/info/headct) : CT scanning uses special x-ray equipment and sophisticated computers to produce multiple pictures of the inside of the body. To better locate and image AVMs, your doctor may perform CT angiography (CTA) (https://www.radiologyinfo.org/en/info/angioct) . CTA uses an injection of contrast material to show the arteries connecting to the AVM or the veins that are draining blood from the AVM . Your doctor may also take images that detect blood flow in the surrounding tissues. This is called CT perfusion

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(CTP) (https://www.radiologyinfo.org/en/info/perfusionheadct) .

Angiography (https://www.radiologyinfo.org/en/info/angiocerebral) : Angiography produces pictures of major blood vessels supplying the AVM. The doctor inserts a thin plastic tube called a catheter into a blood vessel and injects contrast material. The doctor takes x-ray pictures of the AVM and may treat the malformation at the same time.

How are AVMs treated?

AVMs will require treatment based on:

- your symptoms
- the location of the AVM.
- your overall health.

Treatment options include:

- Compression therapy (http://www.radiologyinfo.org) , or wearing a tight-fitting garment on the affected area of the body. The garment helps to reduce swelling in an arm or leg.
- Medications to treat symptoms and complications.
- Sclerotherapy (https://www.radiologyinfo.org/en/info/sclerotherapy) . A doctor injects a solution directly into the malformed blood vessel(s). This causes them to shrink and collapse. You may need several sclerotherapy sessions. Your doctor may surgically remove an AVM following sclerotherapy.
- Embolization (https://www.radiologyinfo.org/en/info/dc-embol) : using image guidance, the doctor inserts and maneuvers a catheter through the body to the malformation. The doctor injects a liquid adhesive through the catheter to seal the vessel. They may also embolize the malformation and surgically remove it.
- Surgery. Your doctor may remove a small AVM in a single location using surgery. If the AVM is large or near important organs and structures in the body, only part of the AVM may be removed. Surgical removal may be an option for small peripheral AVMs and brain AVMs that have bled.

Which test, procedure or treatment is best for me?

- Cerebrovascular Diseases-Aneurysm, Vascular Malformation, and Subarachnoid Hemorrhage (https://www.radiologyinfo.org/en/info/acs-cerebrovascular-diseases)

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