

What Is a Congenital Vascular Malformation?

Congenital vascular malformations (CVMs) are growths or birthmarks made up of blood vessels that have not developed correctly. They occur in 1% of all births. Because they are rare, diagnosing and treating them can be hard.

When the CVM is on the surface of the body, it is fairly easy to manage. Other types of CVMs can be more serious if they involve arteries, veins, and/or **lymphatic vessels** (part of the lymphatic system that helps protect us from infection and

disease). CVMs can develop in any part of the body. They often occur on the hands and feet.

What Are the Different Kinds of CVMs?

Birthmarks

Not all “birthmarks” are the same. Most are superficial and caused by abnormal collections of small blood vessels near the skin. This type of birthmark does not go away, and it maintains its size and appearance. It is not a health threat and does not require treatment. It can be removed with a laser or covered up by tattooing it to match the skin color, if desired.

The other type of birthmark is a **vascular tumor**, or **hemangioma**. At first, it may appear similar to a birthmark on the surface of the skin. It will grow rapidly, then get progressively smaller. Most hemangiomas disappear completely in a few years (normally between ages 2 and 8). They usually do not need treatment unless they interfere with the functioning of the eye, nose, or mouth. Most cases of childhood hemangiomas go away on their own. Parents can speak to a pediatrician if they have questions or concerns about treatment options. Plastic surgery on the birthmark or its remnants is an option when the child is grown.

Venous CVMs

Half of CVMs occur entirely in the veins. There are two basic types of venous CVMs. Some appear as thin-walled lakes where blood collects. They may develop in grapelike clusters. This type usually does not affect the circulation of blood from veins to the heart. However, they can have an abnormal appearance and can interfere with daily activities. They also have the potential to host a type of nonserious blood clot (not the type that travels to the heart or lungs). These CVMs may be worth treating if the mass is large and causes problems, such as interfering with walking.

The other type of venous CVM involves the deeper central veins. These CVMs often interfere with the veins’ function. Segments of major veins may be absent or narrowed. Conversely, some segments may be greatly widened and expanded. This is called a **venous aneurysm**. Treatment depends on how severely the CVM affects blood flow from veins back to the heart, or whether it contributes to **deep vein thrombosis** (DVT), a serious

condition that occurs when a blood clot forms in a vein located deep inside the body. Most venous CVMs involve only short sections of veins and do not require treatment.

Arterial-Venous CVMs

An **arteriovenous fistula** (AVF) is an abnormal connection between an artery and a vein. When a cluster of them occurs, they are called **arteriovenous malformations** (AVMs). These are generally the most serious type of CVM.

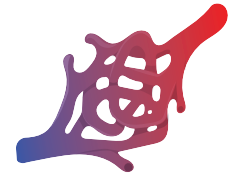
Blood typically flows from your arteries to capillaries, then into the veins. AVF can lead to poor circulation because it causes blood flow to bypass the capillaries and move directly from an artery to a vein. This redirects blood away from tissues and organs. AVFs may also force the heart to work harder.

AVFs are most common in the arms and legs, where they are easiest to manage. When they involve pelvic vessels (in the hip/thigh region), or blood vessels to the brain or vital organs, they can be very difficult to treat. Although AVFs make up only one-third of all CVMs, they receive the most attention because of the serious problems they can create. They are the type of CVMs most likely to require treatment.

NORMAL VESSELS VS. AVM



NORMAL



AVM

Arterial CVMs

CVMs of the arteries are the least common type. They make up only 1 to 2% of all cases. The most common arterial CVM occurs when a segment of an artery fails to develop. This causes blood to flow through an undeveloped side channel, called a **collateral artery**. If the collateral artery is compressed or injured, it could lead to an aneurysm or sudden clotting.

What Are the Symptoms of a CVM?

You can see or feel birthmark-type CVMs on the hands and feet. Varicose veins can be a cause of a CVM, as can blood vessels that injure or bleed easily, or that lead to open wounds (ulcers). CVMs may also cause a limb to become enlarged or grow longer. An AVF may restrict circulation so that tissues and nerves do not get enough blood and oxygen, causing pain.

How Is a CVM Diagnosed?

In the past, the only sure way to evaluate blood vessel problems was by **angiogram**. That is a type of X-ray image taken after contrast dye is injected into the blood vessel. Since most CVMs do not need treatment right away, angiograms are now rarely needed as a first step.

In addition, less invasive imaging is now available. Localized CVMs found on the surface of the skin can often be viewed with a type of ultrasound imaging called a **color duplex scan**. Larger mass lesions can best be studied by **magnetic resonance imaging** (MRI), which creates an image from multiple angles. MRI can help physicians see the



extent of the CVM. An MRI also shows whether the CVM is near muscles, nerves, bones, or joints whose proximity might complicate treatment.

What Are the Treatment Options for CVMs?

CVMs should be treated when they cause problems. Issues could include pain, ulcers, bleeding, blood clots, blockage of major blood vessels, or problems with the way limbs function. The timing of any treatment should be planned according to a child's growth and development. It is often

better to delay operating on very young children, if possible.

In the past, surgery to remove a CVM was the only available treatment. Today, only about 10 to 15% of CVMs are surgically removed. Surgery can be risky—removing even the simplest CVM could lead to significant blood loss.

Surgery may still be needed in some cases, but treatments using catheters have become more common. Catheters are placed into the lesions (usually through a groin vessel), and the abnormal vessels are **embolized**. That means they are blocked

with injectable particles, substances, or devices. This technique can help prevent surgery or can shrink larger CVMs to make them more treatable by surgery.

Laser therapy may also work for small, localized birthmarks. Certain patients, such as those with **Klippel–Trenaunay Syndrome**, a rare venous malformation of the limbs, could benefit from compression garments and bandages. Surgery or less invasive therapy for enlarged superficial veins can also be helpful.

Questions for Your Doctor

It is important to work closely with your health care provider about your diagnosis and treatment. You can bring this form with you to help talk to your medical provider about any questions and concerns you may have.



