Treatment of Vein of Galen Aneurysmal Malformations (VGAM)

What is a Vein of Galen Aneurysmal Malformation (VGAM)?

Under normal conditions, a fine network of blood vessels, called capillaries, delivers oxygen throughout the brain (Figure 1). These capillaries connect the arteries, which carry oxygen-rich blood under pressure from the heart to the brain, and the veins, which return blood to the heart at very low pressure.
A Vein of Galen Aneurysmal Malformation (VGAM) affects the capillary network that drains into the Vein of Galen (Figure 2). These malformed, dilated blood vessels (the aneurysm) create a direct shortcut between arteries and veins in the central part of the brain. This malformation develops before birth, but it is detected in only about half of affected fetuses.
The main consequence is a significant increase in blood flow and pressure in the veins of the malformation and the surrounding brain tissue. After birth, possible symptoms include:

- Neurological dysfunction (transient or permanent deficits, seizures, or developmental delays in children)
- Abnormal increase in head circumference due to excess cerebrospinal fluid
- Heart failure and breathing difficulties, because the heart is overworked by the very high blood flow through the malformation

Without treatment, this malformation often leads to the child's death within months or years.
Approximately 15% of cases have a genetic cause linked to mutations in the EPHB4 gene or Rasa1 gene, and this mutation may be inherited. Other genetic mutations may be investigated if another type of Vein of Galen malformation is suspected. If the appearance of the malformation suggests a genetic origin, genetic screening and counseling may be offered.

What is embolization treatment?

Cerebral angiography (an X-ray examination of the blood vessels) is performed to analyze the structure of the malformation. This examination can also be used to deliver minimally invasive endovascular treatment, called embolization, performed under general anesthesia. During the procedure, a microcatheter (tiny tube) is inserted through the femoral artery in the groin (Figure 3) and advanced into the malformation (Figure 4). A biological glue is then injected to block abnormal blood flow and reduce pressure. Multiple embolization procedures may be necessary depending on the severity and characteristics of the malformation.

What are the possible complications?

While embolization is an effective treatment, rare but potentially serious complications may occur. However, the risks of an untreated VGAM (such as brain hemorrhage or heart failure) are generally much higher than the risks of treatment.

Possible complications include:

- Ischemic stroke (blockage of a brain artery)
- Hemorrhagic stroke (rupture of the malformation, leading to bleeding in the brain)

These complications may result in temporary or permanent disability, such as:

- Paralysis in part of the body
- Speech difficulties
- Vision problems
- Psychomotor delay

In rare cases, these complications may be fatal.

Warning Signs if Treatment Is Delayed

An emergency intervention may be necessary if the following symptoms appear in an infant:

- Abnormal increase in head circumference
- Newly prominent veins on the face
- Unusual irritability, excessive sleepiness, poor head control, or reduced responsiveness
- Difficulty finishing a feeding, excessive sweating while feeding, or poor weight gain

Is embolization covered by health insurance?

Yes, this treatment is covered by basic health insurance (LAMal).

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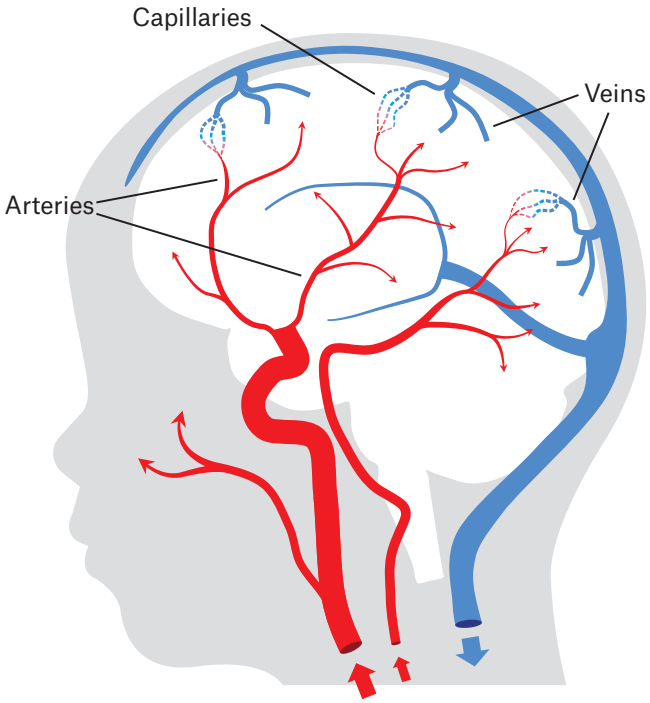
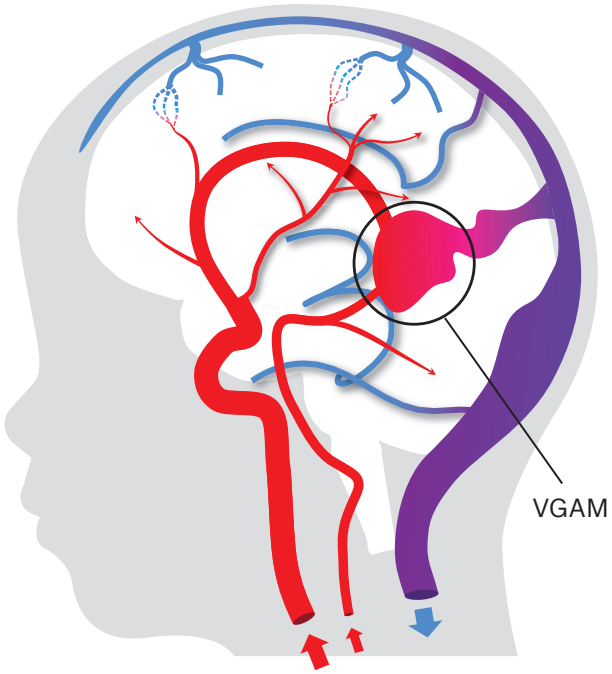


Normal anatomy

1



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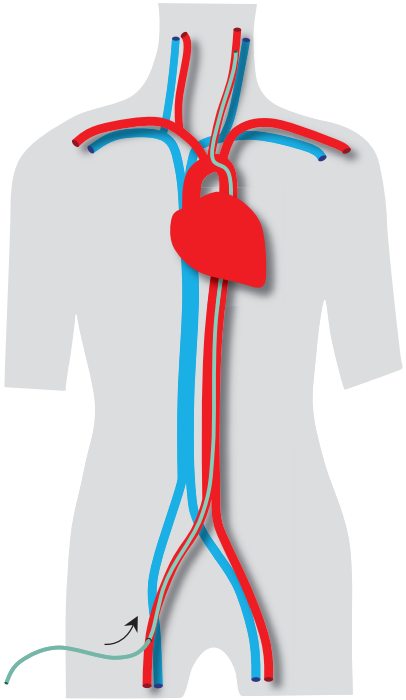
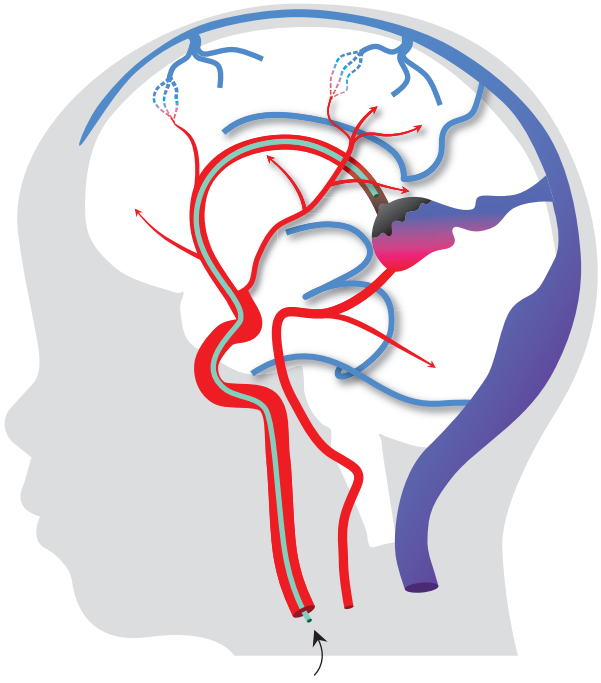


Insertion of a microcatheter by puncture
of the femoral artery or vein in the groin

4



Positioning of the catheter in the malformation
and embolization (occlusion) by injection of biological glue

Contact Information

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