

BRAIN (Cerebral) VASCULAR MALFORMATIONS & HHT



COMPANION FACTSHEET TO
MY HHT CARE CHECKLISTS

SIGNS AND SYMPTOMS

PERSISTENT HEADACHES

SEIZURES

WEAKNESS

NUMBNESS IN THE BODY

VISION CHANGES

SPEECH PROBLEMS

FACTSHEET
FS

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SOME IMPORTANT FACTS TO REMEMBER ABOUT HHT ARE:

At least 10% of people with HHT have brain VMs.

Brain VMs can be life-threatening or disabling if they bleed.

Since brain VMs often do not cause warning symptoms prior to bleeding, screening is recommended in all people with HHT, even infants.

Brain VMs can be successfully treated in most cases.



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Brain (cerebral) vascular malformations (VMs), are abnormal vessels with direct artery to vein connections in the brain. At least 10% of people with **HHT (Hereditary Hemorrhagic Telangiectasia)** have VMs in the brain (cerebral) blood vessels. They are generally thought to arise during embryonic or fetal development. Without treatment, brain VMs are a common cause of **hemorrhagic stroke** (bleeding in the brain) in HHT families. People are often unaware that they have brain VMs until they develop a life-threatening complication, such as **stroke** or **seizure**.

With the right screening and treatment, these life-threatening complications can be prevented, however, there is no single 'standard treatment' that can be recommended for all brain VMs in HHT at the current time.

HERE ARE SOME THINGS TO DISCUSS WITH YOUR PHYSICIAN:

Whether you are due for routine screening for brain VMs.

If you have been previously diagnosed with and/or treated for brain VMs.

If you have any of the listed signs and symptoms.

Getting screening and/or treatment at a HHT Center of Excellence.

HOW IT IS DIAGNOSED

- > **Magnetic Resonance Imaging (MRI):** The recommended test for identifying **brain VMs**. This test utilizes strong magnetic fields to form images of the body. No radiation is used during this study. An **IV** will need to be started for contrast (dye) to be given. The scanner resembles a large tube and the patient is required to lie still during the actual MRI scanning. If the patient has **claustrophobia**, the doctor may prescribe an oral medication to take prior to the MRI. This typically requires **sedation** or anesthesia in young children.
- > **Brain (cerebral) angiogram:** May be recommended if a **brain VM** is identified on MRI. It is a minimally invasive procedure performed by a **neurointerventional radiologist** in an angiography suite. The patient is given **sedation** or general anesthesia for this procedure. A **catheter** (a small tube) is inserted into an **artery** in the top of the thigh and directed through the blood vessels in the body to arteries in the neck or the brain. After the procedure, the patient is observed for several hours or overnight before being discharged home.
- > **Screening** should be performed at the time of initial clinical evaluation for HHT.
- > Patients with **brain VMs** should be referred to a center with neurovascular expertise to be considered for further testing and individualized management.

TREATMENT

- > **Brain (cerebral) embolization:** A procedure performed to block the blood flow to the abnormal vessels. The patient is given **sedation** or general anesthesia for this procedure. In an angiography suite, a **catheter** (a small tube) is inserted into an **artery** in the top of the thigh and directed through the blood vessels in the body to arteries in the brain. An **agent** is then inserted to block off blood flow into the VM and lessen the risk of stroke. After the procedure, the patient is observed overnight before being discharged home.
- > **Surgical Removal:** A surgical procedure to place a clip on the VM or to remove the VM. This procedure is fully curative, but has a higher complication rate than embolization.
- > **Stereotactic Radiosurgery (Gamma Knife):** A type of **focused radiation** that destroys the AVM tissue. This is often done after embolization to ensure that the VM is cured.
- > Oftentimes, treatment requires a combination of the above procedures.
- > Treatment should be performed by a clinician with neurovascular expertise.

AFFILIATED ISSUES

- > Brain hemorrhage
- > Seizures



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