

MY BRAIN VM CARE CHECKLIST

USING THE HHT GUIDELINES

Brain VMs= brain vascular malformations AKA cerebral VMs. The HHT Brain VM Guidelines are detailed on the next pages.

Date: _____

Name: _____

Please check all that apply

I HAVE HHT OR MIGHT HAVE HHT (THE HHT DIAGNOSIS HAS NOT BEEN RULED OUT).

- Talk to my doctor about whether brain VM screening is appropriate for me.
- If I choose to have brain VM screening, ask my doctor for an MRI of the brain, with and without contrast, and also special sequences to detect blood products.
- Consider having my screening done at an HHT Center of Excellence.

I HAVE BRAIN VMs AND I HAVE HAD A BLEED IN MY BRAIN.

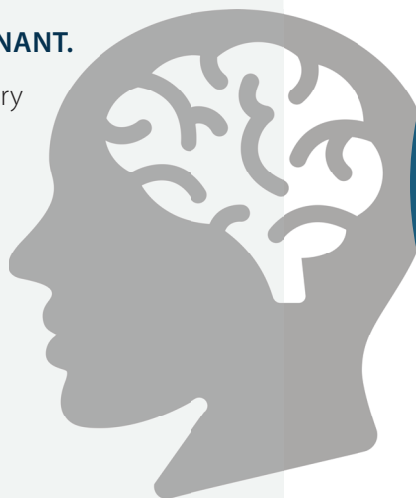
- Ask my doctor to refer me to a center with neurovascular disease, so that I can be considered for definitive treatment of my brain VMs.

I HAVE BRAIN VMs, BUT THEY HAVE NEVER BLED.

- Ask my doctor to refer me to a center with neurovascular disease, so that I can be considered for further testing to confirm the brain VMs.
- Ask my doctor to refer me to a center with neurovascular disease, so that I can get an expert opinion about whether my brain VMs should be treated.

I HAVE BRAIN VMs AND I AM PREGNANT.

- Review the Pregnancy and Delivery HHT Care Checklist.



WHAT ARE THE HHT GUIDELINES AND WHY ARE THEY IMPORTANT?

- The HHT Guidelines are recommendations for care based on evidence and expertise from HHT experts from around the world.
- The HHT Guidelines help ensure that people living with HHT get the best care possible.

WHAT IS MY ROLE AS SOMEONE LIVING WITH HHT?

- Be aware of the Guidelines. Share them with your care team. Ideally you should be seen at an HHT Center of Excellence or your care team may want to consult with one.
- Read up on your condition and know what care is available for HHT.
- **Prepare ahead of time for your appointments:** Bring your HHT Care Checklists and a family member or friend. They can help you communicate your questions and priorities, as well as act as a second set of ears. Share your experiences, worries and priorities to help your care team better understand your needs and provide individualized care.

BRAIN VMs IN HHT

Vascular malformations (VMs), are abnormal vessels with direct artery to vein connections. At least 10% of people with HHT have VMs in the brain (cerebral) blood vessels. 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.



HHT GUIDELINES RECOMMENDATIONS

BRAIN VMs IN HHT

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The expert panel recommends:
(all recommendations are from the First HHT Guidelines)

H1 THE USE OF MRI FOR CEREBRAL VM SCREENING IN ADULTS WITH POSSIBLE OR DEFINITE HHT USING A PROTOCOL WITH AND WITHOUT CONTRAST ADMINISTRATION AND USING SEQUENCES THAT DETECT BLOOD PRODUCTS, TO MAXIMIZE SENSITIVITY.

Clinical Considerations: If patients have received previous embolization, coil compatibility with MRI must be confirmed prior to MR examination. The expert panel acknowledges that the optimum age for adult screening remains unknown but felt that age 18 was appropriate as patients enter adulthood. In the presence of a negative MRI in adulthood no further screening tests are suggested. There may be additional benefits to performing an MRI at initial assessment, in the detection of infarcts and other central nervous system complications of HHT.

H2 THAT ADULTS PRESENTING WITH AN ACUTE HEMORRHAGE SECONDARY TO A CEREBRAL VM BE CONSIDERED FOR DEFINITIVE TREATMENT IN A CENTER WITH NEUROVASCULAR EXPERTISE.

H3 THAT ALL OTHER ADULTS WITH CEREBRAL VMs BE REFERRED TO A CENTER WITH NEUROVASCULAR EXPERTISE TO BE CONSIDERED FOR INVASIVE TESTING AND INDIVIDUALIZED MANAGEMENT.

Clinical Considerations: The expert panel recognizes that asymptomatic cerebral VMs discovered during screening of HHT patients may carry a more favorable natural history. These patients should be managed on an individualized basis. Since some cerebral VMs may carry a favorable natural history, referral to a center with neurovascular expertise prior to performing invasive imaging (catheter angiography) may minimize unnecessary testing.

H4 THAT PREGNANT WOMEN WITH SUSPECTED OR CONFIRMED HHT HARBORING AN ASYMPTOMATIC CEREBRAL VM DURING PREGNANCY HAVE DEFINITIVE TREATMENT OF THEIR CEREBRAL VM DEFERRED UNTIL AFTER DELIVERY OF THEIR FETUS. THE EXPERT PANEL RECOMMENDS THAT THE DELIVERY OF THE FETUS FOLLOW OBSTETRICAL PRINCIPLES.

