MY LIVER VM CARE CHECKLIST

USING THE HHT GUIDELINES

Liver VMs= liver vascular malformations. The HHT Liver VM Guidelines are detailed on the next pages.

Date: _____

Name: _____

Please check all that apply

☐ I AM CONCERNED THAT I MIGHT HAVE LIVER VMs EVEN THOUGH I HAVE NOT HAD ANY SYMPTOMS OR COMPLICATIONS OF LIVER VMs.

- ☐ Talk to my doctor about being screened for liver VMs, and the full range of options available, from "routine physical and bloodwork" to specialized imaging.

☐ I AM CONCERNED THAT I MIGHT HAVE SYMPTOMS OR COMPLICATIONS OF LIVER VMs, SUCH AS HEART FAILURE, PULMONARY HYPERTENSION, CHRONIC LIVER PAIN, BILE DUCT INFECTIONS, ETC.

- ☐ Talk to my doctor about getting diagnostic imaging for liver VMs, using specialized Doppler ultrasound, CT or MRI with special contrast (dye) protocols.
- ☐ Ask my doctor about getting an expert opinion at an HHT Center of Excellence.
- ☐ Ask my doctor about getting a cardiac echo to look for cardiac effects of liver VMs.

☐ I HAVE SYMPTOMS OR COMPLICATIONS FROM LIVER VMs, SUCH AS HEART FAILURE, PULMONARY HYPERTENSION, CHRONIC LIVER PAIN, BILE DUCT INFECTIONS, PORTAL HYPERTENSION, OR OTHER.

- ☐ Ask my doctor about getting an expert opinion at an HHT Center of Excellence.
- ☐ Talk to my doctor about getting first-line treatment of my specific liver VM complications, as detailed in the full HHT Guidelines online supplement.
- ☐ Ask my doctor about involving a cardiologist or pulmonary hypertension doctor with HHT experience if my liver VMs are causing heart problems.
- ☐ Avoid biopsy of the liver.
- ☐ Avoid embolization of liver VMs, in most cases.

☐ I HAVE ONGOING HEART FAILURE SYMPTOMS DESPITE FIRST-LINE MANAGEMENT.

- ☐ See my doctor about treatment with intravenous bevacizumab.
- ☐ See my doctor about consideration for liver transplant.

☐ I HAVE ONGOING BILE DUCT PROBLEMS OR LIVER DYSFUNCTION DESPITE FIRST-LINE MANAGEMENT.

- ☐ See my doctor about consideration for liver transplant.

INTERNATIONAL HHT GUIDELINES

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.

LIVER VMs IN HHT

Liver VMs occur in approximately 75% of HHT patients, more commonly in women and often presenting in the 5th decade. The clinical presentation is typically more severe in patients with ACVRL1 mutation (HHT2). Liver VMs in HHT typically present as diffuse small lesions (telangiectases) throughout the liver, and rarely as discrete large AVMs. Clinicians should offer screening for liver VMs and be aware of possible symptoms or complications and prognostic factors. First-line management depends on symptoms.

HHT GUIDELINES RECOMMENDATIONS

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The expert panel recommends:

D1 **THAT SCREENING FOR LIVER VMs BE OFFERED TO ADULTS WITH DEFINITE OR SUSPECTED HHT.**

Clinical Considerations: The rationale for screening is that awareness of liver VMs could improve subsequent patient management or help confirm the diagnosis of HHT. The imaging test of choice is Doppler ultrasound due to its accuracy, safety, tolerability, low costs and operating characteristics. However, depending on local Doppler ultrasound availability and expertise, as well as patient preference, patients may be screened clinically (history, physical and blood work) or alternative imaging may be considered, including multiphase contrast CT or MRI.

D2 **DIAGNOSTIC TESTING FOR LIVER VMs IN HHT PATIENTS WITH SYMPTOMS AND/OR SIGNS SUGGESTIVE OF COMPLICATED LIVER VMs (INCLUDING HEART FAILURE, PULMONARY HYPERTENSION, ABNORMAL CARDIAC BIOMARKERS, ABNORMAL LIVER FUNCTION TESTS, ABDOMINAL PAIN, PORTAL HYPERTENSION OR ENCEPHALOPATHY), USING DOPPLER ULTRASOUND, MULTIPHASE CONTRAST CT SCAN OR CONTRAST ABDOMINAL MRI FOR DIAGNOSTIC ASSESSMENT OF LIVER VMs.**

Clinical Considerations: The choice of imaging modality should be informed by the risk/benefit balance, local expertise and availability/cost. Contrast studies (CT and MRI) should be avoided if kidney dysfunction. Echocardiography provides additional information about the hemodynamic impact of liver VMs. These tests will be most informative when performed in a center with HHT expertise, in the context of a clinical assessment at an HHT Center of Excellence.

D3 **AN INTENSIVE FIRST-LINE MANAGEMENT ONLY FOR PATIENTS WITH COMPLICATED AND/OR SYMPTOMATIC LIVER VMs, TAILORED TO THE TYPE OF LIVER VM COMPLICATION(S).**

THAT HHT PATIENTS WITH HIGH-OUTPUT CARDIAC FAILURE AND PULMONARY HYPERTENSION BE CO-MANAGED BY THE HHT CENTER OF EXCELLENCE AND AN HHT CARDIOLOGIST OR A PULMONARY HYPERTENSION SPECIALTY CLINIC.

Clinical Considerations: First-line therapies, by specific liver VM complications, are described in the online supplement (see www.HHTGuidelines.org). Typically, patients with symptomatic liver VMs are managed by an expert team at an HHT Center of Excellence, with at least annual follow-up.

D4 **THAT CLINICIANS ESTIMATE PROGNOSIS OF LIVER VMs USING AVAILABLE PREDICTORS, TO IDENTIFY PATIENTS IN NEED OF CLOSER MONITORING.**

Clinical Considerations: Clinicians should plan monitoring for patients with liver VMs patients based on estimated prognosis.

D5 **CONSIDERING INTRAVENOUS BEVACIZUMAB FOR PATIENTS WITH SYMPTOMATIC HIGH-OUTPUT CARDIAC FAILURE DUE TO LIVER VMs WHO HAVE FAILED TO RESPOND SUFFICIENTLY TO FIRST-LINE MANAGEMENT.**

Clinical Considerations: Prescribing and safety monitoring guidance for IV bevacizumab is detailed in Supplement Table 4 (see www.HHTGuidelines.org).

D6 **REFERRAL FOR CONSIDERATION OF LIVER TRANSPLANTATION FOR PATIENTS WITH SYMPTOMATIC COMPLICATIONS OF LIVER VMS, SPECIFICALLY REFRACTORY HIGH-OUTPUT CARDIAC FAILURE, BILIARY ISCHEMIA OR COMPLICATED PORTAL HYPERTENSION.**

Clinical Considerations: Timing for listing a symptomatic patient for orthotopic liver transplantation (OLT) should be based on prognostic predictors and the severity of liver VMs complications, including pulmonary hypertension. Liver transplant can be undertaken in the presence of pulmonary hypertension if pulmonary vascular resistance, estimated by right heart catheterization, is < 3 Woods Units.

From the First HHT Guidelines:

D7 **THAT LIVER BIOPSY BE AVOIDED IN ANY PATIENT WITH PROVEN OR SUSPECTED HHT.**

Clinical Considerations: The rationale for recommending against liver biopsy for diagnosis of liver VMs is that the diagnosis is established with imaging studies whereas biopsy exposes the patient to an unnecessary risk of hemorrhage.

D8 **THAT HEPATIC ARTERY EMBOLIZATION BE AVOIDED IN PATIENTS WITH LIVER VMS AS IT IS ONLY A TEMPORIZING PROCEDURE ASSOCIATED WITH SIGNIFICANT MORBIDITY AND MORTALITY.**

Clinical Considerations: Given the elevated risk of post-embolization necrosis, and death this procedure should not be considered as a first-line therapeutic option. It may be reasonable for the clinician to consider hepatic artery embolization in certain patients, such as a patient with heart failure who has failed to respond to optimal medical therapy and who does not have biliary ischemia or portovenous shunting and who is not a transplant candidate. The risks and benefits of embolization and transplant should be considered on an individualized basis, based on type of shunting, clinical syndrome, patient characteristics and patient preference.