MY PREGNANCY & DELIVERY CARE CHECKLIST

USING THE HHT GUIDELINES

The HHT Pregnancy and Delivery Guidelines are detailed on the next pages

Date: _____

Name: _____

Please check all that apply

☐ I AM EXPECTING A CHILD OR PLANNING A FAMILY, AND I OR MY PARTNER HAVE HHT.

☐ Talk to my doctor about the range of options for genetic diagnosis, from pre-conception to post-delivery.

☐ I HAVE HHT AND I HAVE HAD A PREVIOUS BRAIN HEMORRHAGE OR HAVE OTHER SYMPTOMS THAT SUGGEST I MIGHT HAVE BRAIN VASCULAR MALFORMATIONS (VMs).

☐ Talk to my doctor about planning a brain MRI in second trimester.

☐ I HAVE HHT AND I AM PREGNANT, AND I HAVE NOT RECENTLY BEEN SCREENED FOR LUNG ARTERIOVENOUS MALFORMATIONS (AVMs)

☐ Ask my doctor about planning lung AVM screening tests in second trimester, with bubble echo or non-contrast CT scan, if I am asymptomatic.

☐ Ask my doctor about planning an ASAP diagnostic non-contrast CT scan, if I am having symptoms of lung AVMs.

☐ I HAVE LUNG AVMs AND I AM PREGNANT.

☐ Ask for referral to a center with high-risk pregnancy care and HHT expertise.

☐ Plan to have my lung AVMs treated in early second trimester, at an expert center.

☐ I HAVE BRAIN VMs AND I AM PREGNANT.

☐ Ask for referral to a center with high-risk pregnancy care and HHT expertise.

☐ Talk to an expert multidisciplinary neurovascular team about my brain VMs and my risk of bleeding, to decide if I can proceed with vaginal delivery.

☐ Talk to an expert multidisciplinary team about my brain VMs and my risk of bleeding, to decide if I should have brain VMs treated after delivery.

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.

PREGNANCY & DELIVERY

A pregnant woman with HHT should be assessed for their risk of pregnancy and delivery related complications and have access to, as needed, a multidisciplinary maternal-fetal medicine team that includes HHT experts. Screening for pulmonary AVMs and brain VMs should be considered, as detailed below, and unscreened patients may need to be considered high-risk. In addition, since offspring are at 50% risk of inheriting the pathogenic mutation, pre-pregnancy consultation with an obstetrician is recommended, for consideration of options for genetic diagnosis.

HHT GUIDELINES RECOMMENDATIONS

PREGNANCY & DELIVERY

A pregnant woman with HHT should be assessed for their risk of pregnancy and delivery related complications and have access to, as needed, a multidisciplinary maternal-fetal medicine team that includes HHT experts. Screening for pulmonary AVMs and brain VMs should be considered, as detailed below, and unscreened patients may need to be considered high-risk. In addition, since offspring are at 50% risk of inheriting the pathogenic mutation, pre-pregnancy consultation with an obstetrician is recommended, for consideration of options for genetic diagnosis.

The expert panel recommends:

F1 **THAT CLINICIANS DISCUSS PRE-CONCEPTION AND PRE-NATAL DIAGNOSTIC OPTIONS INCLUDING PRE-IMPLANTATION GENETIC DIAGNOSIS WITH HHT AFFECTED INDIVIDUALS.**

Clinical Considerations: Once the causative familial mutation is identified in an affected parent, then it can be screened for in future off-spring. Available options, including pre-implantation, post-conception and post-delivery testing (online supplement, see www.HHTGuidelines.org), vary internationally. The discussion will be influenced by local legislation pertaining to pre-implantation diagnosis and termination of pregnancy.

F2 **TESTING WITH UNENHANCED MRI IN PREGNANT WOMEN WITH SYMPTOMS SUGGESTIVE OF BRAIN VMs.**

Clinical Considerations: MRI, without gadolinium, should be planned in second trimester, for symptomatic patients including patients with previous cerebral hemorrhage. Asymptomatic patients do not require routine screening during pregnancy.

F3 **THAT PREGNANT WOMEN WITH HHT WHO HAVE NOT BEEN RECENTLY SCREENED AND/OR TREATED FOR PULMONARY AVM SHOULD BE APPROACHED AS FOLLOWS:**

- In asymptomatic patients, initial pulmonary AVM screening should be performed using either agitated saline transthoracic contrast echocardiography (TTCE) or low-dose non-contrast chest CT, depending on local expertise. Chest CT, when performed, should be done early in the second trimester.
- In patients with symptoms suggestive of pulmonary AVM, diagnostic testing should be performed using low-dose non-contrast chest CT. This testing can be performed at any gestational age, as clinically indicated.
- Pulmonary AVMs should be treated starting in the second trimester unless otherwise clinically indicated.

Clinical Considerations: Technique for embolization in pregnant patients should include measures to reduce fetal radiation exposure, including avoidance of fluoroscopy over the abdomen and pelvis, use of pulsed or low-dose fluoroscopy mode, minimizing angiography runs, and use of tight collimation. For both CT and angiography, abdominal shielding is not helpful, and may in fact increase scattered radiation to the fetus.

F4 **THAT PREGNANT WOMEN WITH HHT BE MANAGED AT A TERTIARY CARE CENTER BY A MULTI-DISCIPLINARY TEAM, IF THEY HAVE UNTREATED PULMONARY AVMs AND/OR BRAIN VMs OR HAVE NOT BEEN RECENTLY SCREENED FOR PULMONARY AVMs.**

Clinical Considerations: Pregnant women with untreated pulmonary AVMs or brain VMs, and those who have not been screened, should be considered high risk for hemorrhagic and neurologic complications, and be managed accordingly by a high-risk team with HHT expertise.

F5 NOT WITHHOLDING AN EPIDURAL BECAUSE OF A DIAGNOSIS OF HHT, AND THAT SCREENING FOR SPINAL VASCULAR MALFORMATIONS IS NOT REQUIRED.

Clinical Considerations: Patients should meet with an anesthetist during early third trimester to discuss anesthesia options. The risk of complications from spinal VM during epidural anesthesia are unsubstantiated and only theoretical.

F6 THAT WOMEN WITH KNOWN, NON-HIGH RISK BRAIN VMS CAN LABOR AND PROCEED WITH VAGINAL DELIVERY. PATIENTS MAY REQUIRE AN ASSISTED SECOND STAGE ON A CASE BY CASE BASIS.

Clinical Considerations: If a brain VM has not previously ruptured, patients may proceed with mode of delivery based on obstetrical indications and discussion with their obstetrical care provider. Vaginal delivery is not contra-indicated. Patients with "high risk" brain VMs should be considered for Cesarean section, OR epidural, to allow passive descent of the presenting part, with consideration for an assisted second stage. Diligent management of blood pressure is imperative, in these higher risk cases, and obtaining the opinion a multi-disciplinary neuro vascular team is prudent.