MY GI BLEEDING CARE CHECKLIST

USING THE HHT GUIDELINES

GI Bleeding in HHT is usually chronic bleeding from stomach and/or small bowel. The HHT GI Bleeding Guidelines are detailed on the next pages.

Date: _____

Name: _____

Please check all that apply

☐ I AM CONCERNED THAT I MIGHT HAVE CHRONIC LOW-GRADE BLEEDING FROM TELANGIECTASIA IN MY STOMACH AND/OR BOWELS (GI BLEEDING), BECAUSE I HAVE IRON DEFICIENCY OR ANEMIA.

- ☐ Talk to my doctor about getting a scope of the stomach and small bowel (Upper GI scope AKA Esophagoduodenoscopy or EGD) for diagnosis.
- ☐ Talk to my doctor about getting a capsule endoscopy (camera pill) if my EGD didn't show any significant telangiectasia.
- ☐ Talk to my doctor about whether I am due for a colonoscopy for colon cancer screening.

☐ MY HHT GI BLEEDING IS CAUSING ONGOING IRON DEFICIENCY, DESPITE ORAL IRON SUPPLEMENTS.

- ☐ See my doctor about treatment with tranexamic acid tablets.
- ☐ Avoid routine repetitive endoscopic therapy with Argon Plasma Coagulation, though an initial treatment might be helpful.

☐ MY HHT GI BLEEDING IS CAUSING ONGOING ANEMIA, REQUIRING INTRAVENOUS IRON AND/OR BLOOD TRANSFUSIONS.

- ☐ See an HHT expert about systemic antiangiogenic therapy, such as bevacizumab.

INTERNATIONAL HHT GUIDELINES

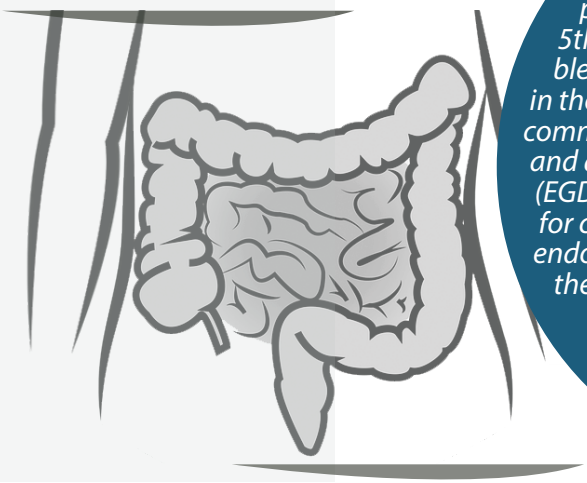
WHAT ARE THE HHT GUIDELINES AND WHY ARE THEY IMPORTANT?

- 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.

GI BLEEDING IN HHT



HHT-related GI bleeding develops in approximately 30% of HHT patients, typically manifesting in the 5th and 6th decades. Chronic low-grade bleeding, typically from GI telangiectases in the stomach and the small bowel, and less commonly in the colon, lead to iron deficiency and anemia. Esophagogastroduodenoscopy (EGD) remains the diagnostic gold standard for confirming HHT-related GI bleeding, but endoscopic management is limited. Medical therapies are often considered, including oral antifibrinolytics and systemic antiangiogenic therapy, with the approach as recommended below.

HHT GUIDELINES RECOMMENDATIONS

GI BLEEDING IN HHT

HHT-related GI bleeding develops in approximately 30% of HHT patients, typically manifesting in the 5th and 6th decades. Chronic low-grade bleeding, typically from GI telangiectases in the stomach and the small bowel, and less commonly in the colon, lead to iron deficiency and anemia. Esophagogastroduodenoscopy (EGD) remains the diagnostic gold standard for confirming HHT-related GI bleeding, but endoscopic management is limited. Medical therapies are often considered, including oral antifibrinolytics and systemic antiangiogenic therapy, with the approach as recommended below.

The expert panel recommends:

B1

ESOPHAGOGASTRODUODENOSCOPY AS THE FIRST LINE DIAGNOSTIC TEST FOR SUSPECTED HHT-RELATED BLEEDING. PATIENTS WHO MEET COLORECTAL CANCER SCREENING CRITERIA AND PATIENTS WITH SMAD4-HHT (GENETICALLY PROVEN OR SUSPECTED) SHOULD ALSO UNDERGO COLONOSCOPY.

Clinical Considerations: Clinicians should consider performing esophagogastroduodenoscopy (EGD) in experienced center, given potential unusual complications during EGD (such as massive epistaxis), and also be aware of precautions required for HHT patients with pulmonary AVMs (Table). In suspected or proven SMAD4-HHT, screening colonoscopy is recommended, starting at age 15 years, repeated every three years if no polyps are found OR every year along with EGD if colonic polyp(s) are found. Other HHT patients (non-SMAD4) should be screened for colon cancer as per general population guidelines.

B2

CONSIDERING CAPSULE ENDOSCOPY FOR SUSPECTED HHT-RELATED BLEEDING, WHEN ESOPHAGOGASTRODUODENOSCOPY DOES NOT REVEAL SIGNIFICANT HHT-RELATED TELANGIECTASIA.

Clinical Considerations: Capsule endoscopy (CE) remains complementary to EGD when anemia is unexplained by the severity of epistaxis and gastric involvement, or when the EGD is negative.

B3

THAT CLINICIANS GRADE THE SEVERITY OF HHT-RELATED GI BLEEDING AND PROPOSES THE FOLLOWING FRAMEWORK:

- Mild HHT-related GI bleeding: Patient who meets their hemoglobin goals* with oral iron replacement.
- Moderate HHT-related GI bleeding: Patient who meets their hemoglobin goals* with IV iron treatment.
- Severe HHT-related GI bleeding: Patient who does not meet their hemoglobin goals* despite adequate iron replacement or requires blood transfusions.

** Hemoglobin goals should reflect age, gender, symptoms and comorbidities.*

Clinical Considerations: Hemoglobin goals (not levels) are specified to reflect the patient's individual physiological needs. This classification applies to patients who have had at least 3 months of iron therapy.

B4

THAT ENDOSCOPIC ARGON PLASMA COAGULATION BE ONLY USED SPARINGLY DURING ENDOSCOPY.

Clinical Considerations: Argon Plasma Coagulation (APC) is best administered concurrent with the initial endoscopic evaluation, for bleeding lesions and significant (1-3 mm) non-bleeding lesions. Repeated APC sessions are discouraged to avoid repeated iatrogenic injury to the intestinal mucosa.

B5 **THAT CLINICIANS CONSIDER TREATMENT OF MILD HHT-RELATED GI BLEEDING WITH ORAL ANTIFIBRINOLYTICS.**

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

B6 **THAT CLINICIANS CONSIDER TREATMENT OF MODERATE TO SEVERE HHT-RELATED GI BLEEDING WITH INTRAVENOUS BEVACIZUMAB OR OTHER SYSTEMIC ANTI-ANGIOGENIC THERAPY.**

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