MY ANEMIA CARE CHECKLIST

USING THE HHT GUIDELINES

The HHT Anemia Guidelines are detailed on the next pages

Date: _____

Name: _____

Please check all that apply

☐ I AM AN ADULT WITH HHT AND SO I AM AT RISK OF ANEMIA AND IRON DEFICIENCY, EVEN IF I HAVE NOT NOTICED ANY BLEEDING.

- ☐ Talk to my doctor about testing for anemia (hemoglobin) and iron deficiency (ferritin).

OR

☐ I AM A CHILD/TEEN WITH HHT WITH SOME BLEEDING SYMPTOMS, AND SO I AM AT RISK OF ANEMIA AND IRON DEFICIENCY.

- ☐ Talk to my doctor about testing for anemia (hemoglobin) and iron deficiency (ferritin).

☐ I HAVE IRON DEFICIENCY OR IRON DEFICIENCY ANEMIA.

- ☐ See my doctor about treatment with oral iron supplements.

☐ I HAVE IRON DEFICIENCY OR IRON DEFICIENCY ANEMIA WHICH HAS NOT IMPROVED ON ORAL IRON SUPPLEMENTS.

- ☐ Talk to my doctor about intravenous iron supplements (also consider this option if I can't tolerate oral iron).
- ☐ Talk to my doctor about additional testing I might need for other causes of anemia.
- ☐ See my doctor about getting blood transfusions if I have severe anemia and I am symptomatic of it, despite intravenous iron, or if I have other serious illness that makes the anemia more dangerous for my health.

☐ I HAVE A MEDICAL INDICATION FOR A BLOOD THINNER (SUCH AS BLOOD CLOT IN A LEG VEIN) OR ANTI-PLATELET THERAPY (SUCH AS HEART DISEASE).

- ☐ Taking one of these medications is not absolutely out of the question; Discuss with an HHT expert as many people with HHT can tolerate.
- ☐ Taking two anti-platelets simultaneously is not absolutely out of the question but is often not tolerated in HHT; discuss with an HHT expert.

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.

ANEMIA & ANTICOAGULATION IN HHT

Anemia is a common complication in people with HHT, occurring in approximately 50%, typically diagnosed in adulthood and only rarely in children with HHT. The primary etiology of anemia is iron deficiency secondary to chronic mucocutaneous bleeding (epistaxis and/or GI bleeding from telangiectases). Patients with HHT should be screened for iron deficiency and anemia, and then supported with iron replacement and red blood cell transfusion, as detailed below. Anticoagulation is not absolutely contraindicated in HHT patients. When there is an indication for anticoagulant or antiplatelet therapy, individualized patient bleeding risks should be considered.

HHT GUIDELINES RECOMMENDATIONS

ANEMIA AND ANTICOAGULATION IN HHT

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The expert panel recommends:

C1

THAT THE FOLLOWING HHT PATIENTS BE TESTED FOR IRON DEFICIENCY AND ANEMIA:

» All adults, regardless of symptoms
» All children with recurrent bleeding and/or symptoms of anemia

Clinical Considerations: Testing typically includes complete blood count (CBC) and ferritin. If anemic but ferritin is not reduced, serum iron, total iron binding capacity, and transferrin saturation should be performed, and a hematology consultation should be considered.

C2

IRON REPLACEMENT FOR TREATMENT OF IRON DEFICIENCY AND ANEMIA AS FOLLOWS:

» Initial therapy with oral iron
» Intravenous iron replacement for patients in whom oral is not effective, not absorbed or not tolerated, or who are presenting with severe anemia

Clinical Considerations: Iron replacement typically starts with once daily oral dosing of 35-65 mg of elemental iron, 2 hours before or 1 hour after meals. An increase in hemoglobin of less than 1.0 gram/dL is considered inadequate in anemic patients, and every-other-day dosing or an alternate oral iron preparation should be attempted. In refractory anemia and/or severe chronic bleeding, regularly scheduled iron infusions may be required. Initial IV iron dosing can be calculated or total initial dose of 1 gram of IV iron can be provided, as single infusion or divided doses. Additional safety and prescribing information are detailed in the online supplement (see link to "Second International Guidelines for the Diagnosis and Management of Hereditary Hemorrhagic Telangiectasia.")

C3

RED BLOOD CELL (RBC) TRANSFUSIONS IN THE FOLLOWING SETTINGS:

» Hemodynamic instability/shock
» Comorbidities that require a higher hemoglobin target
» Need to increase the hemoglobin acutely, such as prior to surgery or during pregnancy
» Inability to maintain an adequate hemoglobin despite frequent iron infusions

Clinical Considerations: Hemoglobin targets and thresholds for RBC transfusion should be individualized in HHT, depending on patient symptoms, severity of ongoing HHT-related bleeding, response to other therapies and iron supplementation, the presence of comorbidities and acuity.

C4 **CONSIDERING EVALUATION FOR ADDITIONAL CAUSES OF ANEMIA IN THE SETTING OF AN INADEQUATE RESPONSE TO IRON REPLACEMENT.**

Clinical Considerations: Evaluation should include measurement of folate, Vitamin B12, MCV, smear, reticulocyte counts, TSH and work-up for hemolysis, with referral to hematology in unresolved cases.

C5 **THAT HHT PATIENTS RECEIVE ANTICOAGULATION (PROPHYLACTIC OR THERAPEUTIC) OR ANTIPLATELET THERAPY WHEN THERE IS AN INDICATION, WITH CONSIDERATION OF THEIR INDIVIDUALIZED BLEEDING RISKS; BLEEDING IN HHT IS NOT AN ABSOLUTE CONTRAINDICATION FOR THESE THERAPIES.**

Clinical Considerations: When anticoagulation is pursued, unfractionated heparin, low molecular weight heparin and vitamin K antagonists are preferred over direct-acting oral anticoagulants, which are less well tolerated in HHT. In cases of atrial fibrillation, if anticoagulation is not tolerated, alternate approaches can be considered, such as left atrial appendage closure.

C6 **AVOIDING THE USE OF DUAL ANTIPLATELET THERAPY AND/OR COMBINATION OF ANTIPLATELET THERAPY AND ANTICOAGULATION, WHERE POSSIBLE, IN PATIENTS WITH HHT.**

Clinical Considerations: If dual or combination therapies are required, duration of therapy should be minimized, and patients should be monitored closely.