

# MY NOSEBLEED (EPISTAXIS) CARE CHECKLIST

USING THE HHT GUIDELINES

The HHT Epistaxis (Nosebleed) Guidelines are detailed on the next pages

Date: \_\_\_\_\_

Name: \_\_\_\_\_

*Please check all that apply*

## I HAVE NOSEBLEEDS (EPISTAXIS) THAT BOTHER ME AND I RATE MY NOSEBLEED SEVERITY TO HELP GUIDE MY NOSEBLEED CARE:

- I rate my usual nosebleeds (circle one) as: mild – moderate -severe.
- My epistaxis severity score (ESS) = \_\_\_\_\_ which is (circle one): mild-moderate-severe.

**My score was computed at this link:**  
<https://www2.drexelmed.edu/HHT-ESS/>

## TALK TO MY DOCTOR ABOUT MOISTURIZING TOPICAL THERAPIES FOR MY NOSE.

## MY NOSEBLEEDS STILL BOTHER ME DESPITE MOISTURIZING TOPICAL THERAPY.

- See my doctor about treatment with tranexamic acid tablets.
- See an HHT-experienced ENT doctor about ablative therapies, like laser, radio frequency, electrosurgery and sclerotherapy.

## MY NOSEBLEEDS ARE AFFECTING MY QUALITY OF LIFE AND/OR ARE LIFE-THREATENING, DESPITE TRANEXAMIC ACID AND/OR ABLATIVE THERAPIES.

- See an HHT expert about systemic antiangiogenic therapy, such as bevacizumab.
- Talk to an HHT-experienced ENT doctor about septodermoplasty surgery.
- Talk to an HHT-experienced ENT doctor about nasal closure surgery (Young's procedure).

## I HAVE A BAD NOSEBLEED AND I AM IN THE EMERGENCY ROOM.

- If I need packing: Ask for packing that won't likely make my nose rebleed, such as liquid packing, dissolvable packing or lubricated low-pressure packing.



## 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.

## EPISTAXIS IN HHT

*Epistaxis is the most common symptom of HHT, developing in 90% of adults with the disease, affecting quality of life and often leading to iron deficiency and anemia. Typically, turbulent nasal airflow with breathing leads to mucosal dryness and bleeding from telangiectases of the nasal mucosa. Topical moisturizing helps prevent the telangiectases from cracking and bleeding and is a mainstay of epistaxis care. When epistaxis doesn't respond to moisturizing, other therapies are considered, including oral antifibrinolytics, ablative therapies, systemic antiangiogenic therapy and surgical management, with the approach as recommended below.*



# HHT GUIDELINES RECOMMENDATIONS

## EPISTAXIS IN HHT

Epistaxis is the most common symptom of HHT, developing in 90% of adults with the disease, affecting quality of life and often leading to iron deficiency and anemia. Typically, turbulent nasal airflow with breathing leads to mucosal dryness and bleeding from telangiectases of the nasal mucosa. Topical moisturizing helps prevent the telangiectases from cracking and bleeding and is a mainstay of epistaxis care. When epistaxis doesn't respond to moisturizing, other therapies are considered, including oral antifibrinolytics, ablative therapies, systemic antiangiogenic therapy and surgical management, with the approach as recommended below.

*The expert panel recommends:*

**A1** PATIENTS WITH HHT-RELATED EPISTAXIS USE MOISTURIZING TOPICAL THERAPIES THAT HUMIDIFY THE NASAL MUCOSA TO REDUCE EPISTAXIS.

*Clinical Considerations:* Topical saline (spray or gel) is typically used twice daily.

**A2** CLINICIANS CONSIDER THE USE OF ORAL TRANEXAMIC ACID FOR THE MANAGEMENT OF EPISTAXIS THAT DOES NOT RESPOND TO MOISTURIZING TOPICAL THERAPIES.

*Clinical Considerations:* Prescribing and safety monitoring guidance for oral tranexamic acid is detailed in Supplement Table 4 (see [www.HHTGuidelines.org](http://www.HHTGuidelines.org))

**A3** CLINICIANS SHOULD CONSIDER ABLATIVE THERAPIES FOR NASAL TELANGIECTASIAS INCLUDING LASER TREATMENT, RADIOFREQUENCY ABLATION, ELECTROSURGERY, AND SCLEROTHERAPY IN PATIENTS THAT HAVE FAILED TO RESPOND TO MOISTURIZING TOPICAL THERAPIES.

*Clinical Considerations:* Clinicians and patients should choose the specific ablative therapy based on local expertise, understanding that ablative therapy is a temporizing treatment for epistaxis and perforation of the nasal septum is a known complication of all techniques.

**A4** CLINICIANS CONSIDER THE USE OF SYSTEMIC ANTIANGIOGENIC AGENTS FOR THE MANAGEMENT OF EPISTAXIS THAT HAS FAILED TO RESPOND TO MOISTURIZING TOPICAL THERAPIES, ABLATIVE THERAPIES AND/OR TRANEXAMIC ACID.

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

**A5** CLINICIANS CONSIDER A SEPTODERMOPLASTY FOR PATIENTS WHOSE EPISTAXIS HAS FAILED TO RESPOND SUFFICIENTLY TO MOISTURIZING TOPICAL THERAPIES, ABLATIVE THERAPIES, AND/OR TRANEXAMIC ACID.

*Clinical Considerations:* Clinicians and patients should consider septodermoplasty when epistaxis affects QOL or is life-threatening, considering risks and benefits, as well as alternatives, such as nasal closure and anti-angiogenic medications.



**A6** CLINICIANS CONSIDER A NASAL CLOSURE FOR PATIENTS WHOSE EPISTAXIS HAS FAILED TO RESPOND SUFFICIENTLY TO MOISTURIZING TOPICAL THERAPIES, ABLATIVE THERAPIES, AND/OR TRANEXAMIC ACID.

*Clinical Considerations:* Clinicians and patients should consider nasal closure when epistaxis affects QOL or is life-threatening, considering risks and benefits, as well as alternatives, such as septodermoplasty and anti-angiogenic medications.

*From the First HHT Guidelines:*

**A7** PHYSICIANS ADVISE PATIENTS WITH HHT-RELATED EPISTAXIS TO USE AGENTS THAT HUMIDIFY THE NASAL MUCOSA TO PREVENT EPISTAXIS.

**A8** CLINICIANS REFER HHT PATIENTS WITH EPISTAXIS AND WHO DESIRE TREATMENT TO OTORHINOLARYNGOLOGISTS WITH HHT EXPERTISE FOR EVALUATION AND TREATMENT.

*Clinical Considerations:* Primary physicians are key players in the care of HHT patients, especially in the emergency situation. In the patient with epistaxis problematic enough to warrant consideration of treatment, consultation with an otorhinolaryngologist with HHT expertise should help guide the intervention choice, to maximize effectiveness and reduce risk, in this life-long rare disorder.

**A9** WHEN CONSIDERING NASAL SURGERY FOR REASONS OTHER THAN EPISTAXIS, THE PATIENT AND CLINICIAN OBTAIN CONSULTATION FROM AN OTORHINOLARYNGOLOGISTS WITH EXPERTISE IN HHT-RELATED EPISTAXIS.

*Clinical Considerations:* In the patient with HHT and an unrelated ENT problem requiring surgery, consultation with an otorhinolaryngologist with HHT expertise should help guide the procedural interventions to minimize risk of worsening epistaxis.

**A10** THE TREATMENT FOR ACUTE EPISTAXIS REQUIRING INTERVENTION INCLUDE PACKING WITH MATERIAL OR PRODUCTS THAT HAVE A LOW LIKELIHOOD OF CAUSING RE-BLEEDING WITH REMOVAL (E.G., LUBRICATED LOW-PRESSURE PNEUMATIC PACKING).

*Clinical Considerations:* In order to perform atraumatic packing, the clinician can lubricate the packing or use a pneumatic packing which allows insertion and removal of the packing in a deflated size. When using pneumatic packing, a low pressure packing would be preferable. This recommendation is specifically addressing nasal packing performed by physicians, though the expert panel is aware that patients often choose to self-pack the nose.