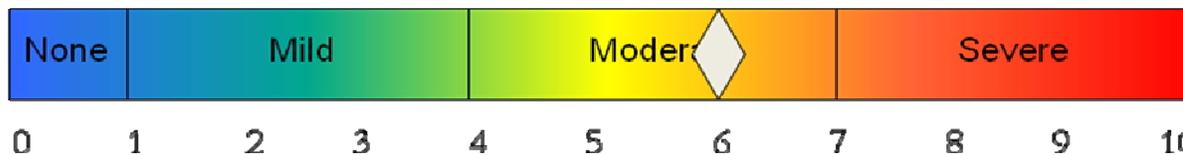


The following scenarios are examples of how individuals should respond to questions from the HHT-ESS. The cases are fictional, but are designed to highlight some of the possible areas of confusion related to the questions in the severity score.

**Example 1.** A 55 year old woman with a history of HHT seeks medical attention in an Emergency Room with nose bleeding that would not stop. In the Emergency Room, she is treated with nasal packing. Her recent nose bleeds usually last about 20 minutes in duration, and she experiences several bleeds each week. She has low blood counts for which she has been taking oral iron pills, but she has not received a blood transfusion in the past. Although she had this recent extreme nose bleed, her average bleeding requires several tissues.

1. How often do you TYPICALLY have nose bleeding?
  - Several per week
2. How long do your TYPICALLY nose bleeding last?
  - 16-30 minutes
3. How would you describe your TYPICAL nose bleeding intensity?
  - Not Typically Gushing or Pouring
4. Have you sought medical attention for your nose bleeding?
  - Yes
5. Are you anemic (low blood counts) currently?
  - Yes
6. Have you received a red blood cell transfusion SPECIFICALLY for nose bleeding?
  - No

**Normalized ESS = 5.98**

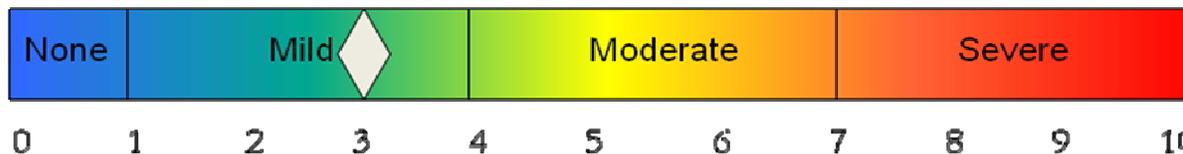


**Example 2.** A 31 year old man with HHT who has not seen a physician related to nose bleeding is interested in comparing his nose bleeding severity

with others in his family. He has some nose bleeding each day, but the bleeding lasts only between 1-2 minutes. His bleeding intensity is a slow drip. He does not go to the doctor, so he is not sure if he is anemic, but does not think he is. He has never received a blood transfusion.

1. How often do you TYPICALLY have nose bleeding?
  - Once per day
2. How long do your TYPICALLY nose bleeding last?
  - 1-5 minutes
3. How would you describe your TYPICAL nose bleeding intensity?
  - Not Typically Gushing or Pouring
4. Have you sought medical attention for your nose bleeding?
  - No
5. Are you anemic (low blood counts) currently?
  - No
6. Have you received a red blood cell transfusion SPECIFICALLY for nose bleeding?
  - No

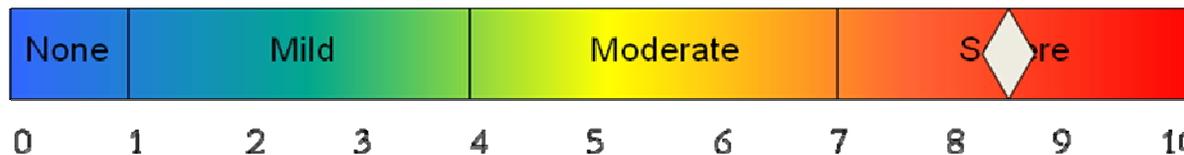
**Normalized ESS = 3.02**



**Example 3.** A 44 year old woman with HHT has scheduled an appointment with one of the HHT Centers of Excellence because her nose bleeding has been worsening. She would like to come to this appointment with more information for the Center, so she decided to complete the Epistaxis Severity Score on-line before her appointment. She has seen a local ENT (Otolaryngologist) who has performed multiple procedures to try to control her nose bleeding. She usually has one nose bleed per week, but the episode lasts more than an hour and blood pours out with each episode. She has received intravenous iron for anemia and has had many blood transfusions for nose bleeding.

1. How often do you TYPICALLY have nose bleeding?
  - Once per week
2. How long do your TYPICALLY nose bleeding last?
  - >30 minutes
3. How would you describe your TYPICAL nose bleeding intensity?
  - Typically Gushing or Pouring
4. Have you sought medical attention for your nose bleeding?
  - Yes
5. Are you anemic (low blood counts) currently?
  - Yes
6. Have you received a red blood cell transfusion SPECIFICALLY for nose bleeding?
  - Yes

**Normalized ESS = 8.40**



**Other Considerations:**

- Questions 1-3 should be reflective of TYPICAL (or average) symptoms of nose bleeding. One-An occasional bad episode (frequency, duration or intensity) should not influence how TYPICAL symptoms are scored.
- Seeking Medical Attention (Question 4) should be positive (“Yes”) if nose bleeding was the SPECIFIC reason medical attention was sought. Seeing a physician for a blood pressure check and discussing nose bleeding should not prompt a “Yes” response to this question. GIVE EXAMPLES HERE. For use in clinical trials, the clinical trial visit should NOT count as seeking medical attention unless the individual did not seek an alternative medical care provider because the clinical trial visit was scheduled/planned/more convenient (e.g. “I would have gone to the ER if I wasn’t coming to see the clinical trial physician”).

A LITTLE CONFUSING HERE. Routine clinical trial follow-up visits are not considered seeking medical attention for the purposes of the severity score.

- Anemia (Question 5) should be answered to the best of the individual's knowledge at the time the questionnaire is taken. If information comes available that the individual's blood count would change the answer to this question in a reasonable time from the time the questionnaire was taken, the response should be changed to the appropriate response. For example, a patient fills out the HHT-ESS on the day of a clinic appointment and answers "No" to Question 5. Blood is drawn the following day that demonstrates a hemoglobin level of 9mg/dl (low). The HHT-ESS should be re-scored with Question 5 being changed to "Yes".
- For clinical trials, the time interval for the baseline score should be consistent with the interval of study during the trial. For example, if a study plans to have an HHT-ESS measured every 3 months after an intervention (e.g. laser therapy to the nose), then the baseline HHT-ESS should reflect the participant's symptoms for the 3 months leading into the trial. In this case "SINCE YOUR LAST TREATMENT" should be changed to "IN THE LAST 3 MONTHS". All questions should be considered in the context of this time interval. For Example, Question 1. How often do you TYPICALLY have nose bleeding *in the last 3 months*? Question 4. Have you sought medical attention for your nose bleeding *in the last 3 months*?
- Because this Epistaxis Severity Score is early in clinical use, care providers from the HHT Centers of Excellence are still trying to determine the clinical significance of a particular score (e.g. Normalized Score 5.5) in regards to specific evaluation or treatment recommendations. As more familiarity with the HHT-ESS, the Foundation will provide more detailed recommendation information.