

Other Considerations Concerning the HHT-ESS

1. Questions 1-3 should be reflective of TYPICAL (or average) symptoms of nose bleeding. One bad episode (frequency, duration or intensity) should not influence how TYPICAL symptoms are scored.
2. 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. Seeking medical attention may include primary care physicians, otolaryngologists, hematologist, HHT Center experts, or any other medical provider who may provide care for nose bleeding. 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”). Routine clinical trial follow-up visits are not considered seeking medical attention for the purposes of the severity score.
3. 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”.
4. 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 2 months after an intervention (e.g. laser therapy to the nose), then the baseline HHT-ESS should reflect the participant's symptoms for the 2 months leading into the trial. In this case "DURING THE PAST 3 MONTHS" should be changed to "DURING THE PAST 2 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 2 months*? Question 4. Have you sought medical attention for your nose bleeding *in the last 2 months*?

5. 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.