

SUGGESTED GUIDELINES FOR USE OF THE HHT-ESS IN THE RESEARCH SETTING.

1. Modify the time scale as needed:
 - a. During the past 2 months.
 - b. During the past 3 months.
 - c. During the past 6 months.
 - d. Since the last research visit.
 - e. etcetera
2. Identical time scale modifications should be made in the introductory sentence, and in all questions except number 5.
3. You will need to decide ahead of time how do deal with the circumstance when the patient's answer to Question 5 is different than measurement of the hemoglobin (if done) on the day the patient fills out the ESS questionnaire (e.g., the patient answers no to Question 5, and the measured hemoglobin on that study visit is 10 gm/l). We recommend changing the answer to reflect the actual value obtained.
4. A template for use in research studies is shown below.
5. Calculation of the ESS is possible on the website. For manual calculation, the formula is listed below.

Calculation of the ESS:

Question	Response	Multiplied by:		Coefficient	Result
1	Less than monthly	0	x	0.14	
	Once per month	1			
	Once per week	2			
	Several per week	3			
	Once per day	4			
Several per day	5				
2	< 1 minute	0	x	0.25	
	1-5 minutes	1			
	6-15 minutes	2			
	16-30 minutes	3			
	> 30 minutes	4			
3	No	0	x	0.25	
	Yes	1			
4	No	0	x	0.30	
	Yes	1			
5	No	0	x	0.20	
	Yes	1			
	I Don't Know	Drop			
6	No	0	x	0.31	
	Yes	1			
TOTAL =				Denominator	Raw Score

$$\text{Normalized HHT-ESS} = \left[\frac{\text{Raw Score}}{\text{Denominator}} \right] \times 10$$

Epistaxis Severity Score (ESS) for Hereditary Hemorrhagic Telangiectasia

The purpose of these questions is to calculate a severity score of epistaxis (nose bleeding) for patients with Hereditary Hemorrhagic Telangiectasia who are involved in a Research Study. For more information regarding the meaning of this score, please discuss this with your health care provider.

Please answer each of the following questions as they pertain to your TYPICAL nosebleed symptoms DURING THE PAST 2 MONTHS. Please answer all questions.

1. How often did you TYPICALLY have nosebleeds during the past 2 months?
 - Less than monthly
 - One to three times per month
 - Once per week
 - Several per week
 - Once per day
 - Several per day
2. How long did each nosebleed TYPICALLY last for you during the past 2 months?
 - < 1 minute
 - 1-5 minutes
 - 6-15 minutes
 - 16-30 minutes
 - > 30 minutes
3. How would you describe your TYPICAL nosebleed intensity during the past 2 months?
 - Not Typically Gushing or Pouring
 - Typically Gushing or Pouring
4. Have you sought medical attention OUTSIDE OF THIS RESEARCH STUDY for your nosebleeds during the past 2 months?
 - No
 - Yes
5. Are you anemic (low blood counts) currently?
 - No
 - Yes
6. Have you received a red blood cell transfusion SPECIFICALLY for nosebleeds during the past 2 months?
 - No
 - Yes

Raw Epistaxis Severity Score: _____

Normalized Epistaxis Severity Score: _____