

# Development of a Scoring System for Epistaxis Severity in Hereditary Hemorrhagic Telangiectasia

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**Introduction:** Hereditary Hemorrhagic Telangiectasia-related epistaxis leads to dramatic alterations in social functioning and quality of life. Although more than 95% of patients experience epistaxis, there is considerable heterogeneity in the severity of epistaxis experienced between individuals. Several methods to categorize epistaxis severity have been utilized by care centers; however, little uniformity currently exists among different centers. Because no standardized method exists to categorize epistaxis severity, the purpose of this study was to determine factors associated with patient reported severity in order to develop an epistaxis severity score.

**Methods:** In order to determine factors believed to be associated with epistaxis, HHT care providers and a focused group of patients were interviewed to determine a comprehensive list of possibly associated factors. From this list, an electronic survey was developed and administered to patients with HHT. Descriptive analyses were performed with calculations of means and medians for continuous variables and proportions for categorical variables. Multiple ordinal logistic models were developed to determine risk factors for epistaxis severity. Subsequently, multiple linear regression models were created to develop the epistaxis severity score. Finally, boot-strap sampling methods were employed to assist with model estimation.

**Results:** 914 respondents from 21 countries completed the electronic survey. From this, a cohort of 877 (96%) subjects reported epistaxis. The mean (SD) age was 52.8 (12.9) years and 61.2% were female. Independently associated risk factors for self reported epistaxis severity included frequency of epistaxis (OR 1.56), average duration of bleeding episodes (OR 2.20), intensity of bleeding (OR 2.26), having sought medical attention for epistaxis (OR 2.34), need for epistaxis-specific blood transfusion (OR 3.06), and presence of anemia (OR 1.50,  $p < 0.001$  for all). Using multiple linear regressions, predictors of self reported epistaxis severity are shown in the table.

Variable	Coefficient	Standard Error	p-value
Intensity	0.26	0.07	< 0.001
Frequency	0.15	0.02	< 0.001
Duration	0.25	0.02	< 0.001
Medical Attention	0.32	0.07	< 0.001
Transfusion	0.35	0.06	< 0.001
Anemia	0.13	0.03	< 0.001

Using the coefficients from the linear regression model, an epistaxis severity score was generated. For ease of use, the raw epistaxis severity score was normalized to a range of 0 (none) to 10 (most severe).

Conclusion: Through evaluation of a comprehensive list of possibly related predictors of epistaxis severity obtained from patients and care providers, six factors were utilized to develop an epistaxis severity score. These include: frequency of bleeding episodes, average bleeding duration, intensity of average bleeding episodes, seeking medical attention for nose bleeding, the presence of anemia, and the need for blood transfusion specifically related to epistaxis. We believe that this epistaxis severity score may serve as an outcome measure for assessing clinical status and therapeutic efficacy.

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