

Severity and Features of Epistaxis in Children with a Mucocutaneous Bleeding Disorder

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Objective To use standardized bleeding questionnaires to compare the severity and patterns of epistaxis in children with a mucocutaneous bleeding disorder and control children.

Study design The epistaxis sections of the Pediatric Bleeding Questionnaire (PBQ) administered to pediatric patients with von Willebrand disease or a platelet function disorder and healthy control children were reviewed. Scores and features of epistaxis (frequency, duration, onset, site, seasonal correlation, and need for medical/surgical intervention) were recorded. A PBQ epistaxis score ≥ 2 was defined as clinically significant. The Katsanis epistaxis scoring system was administered to eligible patients, ie, with ≥ 5 episodes of epistaxis per year.

Results PBQ epistaxis scores were obtained for 66 patients, median age 12 years (range 0.6-18.3 years), and 56 control children. The median PBQ epistaxis score in patients was 2 vs 0 in control children ($P < .0001$). All of the features of epistaxis, except spontaneous onset, occurred in a significantly greater proportion of patients than control children with epistaxis. A total of 50% of the patients were graded as having severe epistaxis by the Katsanis epistaxis scoring system, and 30 of these (91%) had a clinically significant PBQ epistaxis score.

Conclusion Standardized bleeding questionnaires are useful in the assessment of epistaxis severity and pattern and may help to distinguish children with and without a mucocutaneous bleeding disorder. (*J Pediatr* 2017;■■■:■■-■■■).

Epistaxis is a common bleeding symptom in children with an inherited mucocutaneous bleeding disorder such as von Willebrand disease (VWD) or a platelet function disorder (PFD) but also occurs in healthy children.¹⁻³ Because of their young age, children generally lack exposure to hemostatic challenges and therefore may not manifest other bleeding symptoms, even when they have a bleeding disorder. Determining the features of epistaxis that are suggestive of an underlying hemostatic defect may help to determine which children presenting with epistaxis should undergo laboratory testing for an inherited bleeding disorder. A standardized approach to the assessment of epistaxis would be useful in this regard and currently is lacking.

To standardize bleeding histories, bleeding assessment tools (BATs) have been developed that aim to discriminate between individuals with and without a bleeding disorder.⁴ Most BATs use a scoring system that scores various mucocutaneous bleeding symptoms based on the medical treatment of the most severe episode of the specific symptom. The overall bleeding score is determined by summing the scores for all bleeding symptoms. In 2009, a pediatric adaptation of a standardized adult bleeding questionnaire and bleeding score, known as the Pediatric Bleeding Questionnaire (PBQ), was validated for use in children with VWD.⁵ The International Society on Thrombosis and Haemostasis (ISTH)-BAT was published in 2010 for use in adults and children.⁶ Both the PBQ and ISTH-BAT assess the severity of various mucocutaneous bleeding symptoms including epistaxis.

Over time, an interest in organ-specific bleeding scores has arisen.⁴ For example, a screening tool for selecting female patients with menorrhagia for hemostatic evaluation has been developed.⁷⁻⁹ Organ-specific information can provide important information, especially in young children in whom epistaxis may be the only bleeding symptom. In 1988, Katsanis et al published a semiquantitative epistaxis scoring

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Ag	Antigen
BAT	Bleeding assessment tool
ESS	Epistaxis scoring system
ISTH	International Society on Thrombosis and Haemostasis
PBQ	Pediatric Bleeding Questionnaire
PFD	Platelet function disorder
RCo	Ristocetin cofactor activity
SickKids	The Hospital for Sick Children (HSC)
VWD	von Willebrand disease
VWF	von Willebrand factor

system (ESS) to assess the severity of recurrent epistaxis, defined as ≥ 5 episodes per year, in children.¹⁰ The authors concluded that their scoring system was clinically useful for both the assessment of epistaxis severity and defining the group of patients requiring further hemostatic testing. To date, the Katsanis ESS has not been validated. For the assessment of the severity of epistaxis, an epistaxis score also can be extracted from the PBQ and ISTH-BAT. Specific features of epistaxis are recorded in these latter questionnaires as well.

The aim of this study was to determine the severity and features of epistaxis in children with VWD or a PFD by using the PBQ and Katsanis ESS compared with healthy children to identify which specific features are discriminatory in identifying an underlying bleeding disorder.

Methods

Patients were children ≤ 18 years of age with a known diagnosis of VWD or a PFD at The Hospital for Sick Children (SickKids) who had been recruited previously for a study of PBQ total bleeding scores.^{11,12} Patient recruitment for the study and laboratory diagnostic criteria for subtypes of VWD and PFDs have been described previously.^{11,12} In particular, patients with type 1 VWD were classified as having definite or possible type 1 VWD. Criteria for the diagnosis of definite type 1 VWD were a von Willebrand factor (VWF) ristocetin cofactor activity (VWF:RCo) of 0.05-0.50 U mL⁻¹ on at least 2 occasions and a VWF antigen (VWF:Ag) of 0.05-0.50 U mL⁻¹ on at least 1 occasion, a ratio of VWF:RCo/VWF:Ag of >0.50 , multimer analysis showing normal or globally reduced VWF multimers, and a positive bleeding history according to SickKids (HSC) criteria.¹³ Possible type 1 VWD referred to patients who had (1) laboratory data that fit the aforementioned criteria but with a negative bleeding history according to SickKids criteria or (2) abnormal VWF:RCo and VWF:Ag on at least 1 occasion with or without a bleeding history. We recognize that there is a possible classification bias in the inclusion of patients in the possible type 1 VWD category with borderline VWF levels on only 1 occasion and without a bleeding history (3/22 in this study), but because there was a suspicion of type 1 VWD in these patients, they could not be classified as unaffected. Diagnosis of a PFD was made according to criteria defined by the Rare Inherited Bleeding Disorders Committee of the Association of Hemophilia Centre Directors of Canada (<https://www.ahcdc.ca/rare-inherited-bleeding-disorders>). The study was approved by the Research Ethics Board at SickKids, and informed consent was obtained from all participants.

Control subjects were healthy children previously recruited to determine the normal range of the PBQ score⁵ and unaffected siblings identified from patients' records as siblings of children with confirmed VWD who had had normal laboratory testing on at least 1 occasion.¹¹ The study was approved by the Institutional Review Board of the Children's Hospital of Oakland and the Research Ethics Board at SickKids; informed consent was obtained from all participants.

Data Collection

The method of PBQ administration has been described by Biss et al.¹¹ Data were extracted from the PBQ epistaxis section of the questionnaire (Figure 1, A; available at www.jpeds.com). This section opens with the question by the interviewer: "Have you ever had a problem with nosebleeds?" Subjects who answered "yes" to this question were considered to have epistaxis. A problem with nosebleeds was further defined as either being "significant" when lasting for >10 minutes or occurring >5 times per year, or "trivial" when the nosebleeds did not meet the criteria for being significant. Scoring of trivial and significant epistaxis is described in the following paragraph. Data extracted from the epistaxis section of the PBQ included the number of epistaxis episodes occurring per year, duration of an average single episode, whether epistaxis was of spontaneous onset (refers to a nosebleed occurring without trauma or nose picking) or occurred in relation to drug ingestion (within 7 days of taking aspirin, aspirin-containing preparations, or other anti-inflammatory medication), if bleeding only ever occurred from one nostril or from both nostrils (either at the same time or at separate times), and the presence of seasonal correlation (defined as epistaxis occurring only during 1 or 2 specified seasons of the year).

For the most severe episode, whether medical attention was required was recorded, as well as the type of medical attention: consultation with a healthcare professional only, packing, cauterization, antifibrinolytics, desmopressin, replacement therapy, or blood transfusion. Each patient or control with epistaxis was graded on a PBQ scale of 0-4 (Figure 1, B; available at www.jpeds.com), depending on clinical severity of the most severe epistaxis episode (0: trivial; 1: >5 per year or >10 minutes duration; 2: consultation with a healthcare professional; 3: packing, cauterization, or antifibrinolytics; 4: blood transfusion, replacement therapy, or desmopressin). Replacement therapy included the administration of 1 or more of the following products to treat epistaxis: platelet transfusions, recombinant factor VIIa, or a VWF-containing concentrate. Clinically significant epistaxis was defined as a PBQ epistaxis score of ≥ 2 . The PBQ and ISTH-BAT epistaxis scoring keys are identical.

Patients with recurrent epistaxis (defined as having ≥ 5 episodes per year) were eligible for grading, and those who had <5 episodes of epistaxis per year were termed "Katsanis ESS ineligible." The Katsanis ESS was administered at the same time as the PBQ by interview of parents of patients/patients by a physician, a research nurse, or a research associate working in the bleeding disorders/pediatric hematology clinic. Epistaxis data extracted from the Katsanis ESS questionnaire included frequency, duration, amount (estimation of average blood loss per episode), epistaxis history/age (eg, the proportion of child's life that nosebleeds had been recurrent), and site (unilateral or bilateral), with each of these features yielding a score from 0 to 2 based on increasing severity. A summative score was then used to classify patients as having mild epistaxis (score 0-6) or severe epistaxis (score 7-10) (Figure 1, C; available at www.jpeds.com). The Katsanis ESS was not administered to the control subjects in this study.

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