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 2018;193:183-9).*

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

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

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bleeding symptom. In 1988, Katsanis et al published a semiquantitative epistaxis scoring 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 score[5] 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.

Table I. Characteristics of control subjects (n = 163) and patients (n = 107)

Diagnoses	Total number of children	Number of children with epistaxis (%)	Male/female ratio	Median age, y (range)	Median PBQ epistaxis score (range)
Control children	163	56 (34)	28:28	12.0 (1.5-17.0)	0 (0-2)
Patients	107	66 (62)	39:27	12.0 (0.6-18.3)	2 (0-4)
Type 1 VWD	63	34 (54)	19:15	12.6 (3.5-17.8)	1.5 (0-4)
Definite	37	23 (62)	12:11	12.6 (3.5-17.8)	2 (0-4)
Possible	26	11 (42)	7:4	12.5 (4.2-16.6)	0 (0-4)
Type 2 VWD*	5	3 (60)	2:1	5.0 (5.0-13.7)	2 (0-4)
Type 3 VWD	16	12 (75)	8:4	6.8 (1.8-16.7)	4 (0-4)
PFD†	23	17 (74)	10:7	10.3 (0.6-18.3)	2 (0-4)

*Patients with type 2A (n = 1), 2B (n = 1), 2M (n = 2), and 2N (n = 1) VWD.
†Patients with Glanzmann thrombasthenia (n = 4), dense granule defects (n = 7), Hermansky-Pudlak syndrome (n = 2), MYH9-related disease (n = 3), Ehlers-Danlos syndrome (n = 2), Noonan syndrome (n = 2), and an unspecified PFD (n = 3).[12]

Statistical Analyses

Kruskal-Wallis 1-way ANOVA was used to compare PBQ epistaxis scores between control subjects, 4 VWD groups (possible type 1, definite type 1, type 2, and type 3), and patients with a PFD. Generalized linear models were used to determine the effect of age and sex on PBQ epistaxis scores. The χ^2 distribution was used to compare features of epistaxis and epistaxis severity between patients and control groups. *P* values < .05 were considered statistically significant. Analysis was done with SAS 9.1 software (SAS Institute Inc, Cary, North Carolina).

Results

A review was done of the epistaxis sections of 107 PBQs administered to a cohort of patients/parents of patients with a known diagnosis of VWD or a PFD, as reported previously.[11,12] Control subjects were 142 healthy children and 21 unaffected siblings of patients with VWD,[5,11] and the epistaxis sections of PBQs administered to the control subjects were reviewed as well. A total of 122 children reported a problem with epistaxis—66 patients with VWD or a PFD and 56 control children (**Table I**). The median age of the patients with epistaxis (59% male) as well as the control subjects with epistaxis (50% male) was 12 years. Only 4 of 66 (6%) patients had epistaxis as their only bleeding symptom, in contrast to 37 of 56 (66%) control children.

The PBQ epistaxis score varied according to patient diagnosis (**Figure 2**). A total of 38 of 66 patients (58%) had a clinically significant PBQ epistaxis score of ≥2, including the majority of patients with definite type 1 VWD (65%), type 3 VWD (75%), and a PFD (59%). The majority of control children (68%) had a PBQ epistaxis score of 0, with only 5 of 56 (9%) having a clinically significant PBQ epistaxis score. The median PBQ epistaxis score was significantly greater in patients than in control subjects (2, range 0-4 vs 0, range 0-2, *P* < .0001).

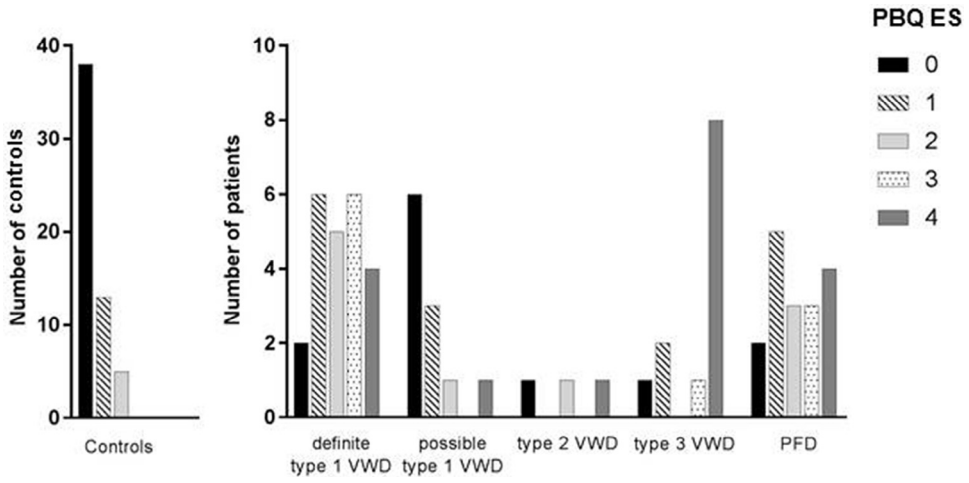


Figure 2. PBQ ES in control children (n = 56) and children with VWD or a PFD (n = 107). *ES*, epistaxis score.

Table II. Features of epistaxis in control subjects (n = 56) and patients (n = 66) with epistaxis

Features of epistaxis	Number of control subjects (%)	Number of patients (%)	*P* value
Epistaxis requiring medical attention*	5/56 (9)	38/66 (58)	<.0001
Consultation with a healthcare professional only	5/5 (100)	10/38 (26)	
Nasal packing, cauterization, or antifibrinolytics	0/5 (0)	10/38 (26)	
Blood transfusion, replacement therapy, or desmopressin	0/5 (0)	18/38 (47)	
Frequency, >5 episodes per year	11/56 (20)	44/66 (67)	<.0001
Duration >10 min	5/56 (9)	41/66 (62)	<.0001
Spontaneous onset	44/56 (79)	57/66 (86)	.23
Occurring from both nostrils	15/56 (27)	51/66 (77)	<.0001
Presence of seasonal correlation	24/56 (43)	14/66 (21)	.022

*If epistaxis required medical attention, the PBQ epistaxis score was ≥2, eg clinically significant (Figure 1, A and B).

Of all the patients who required medical attention, 47% received blood transfusion, desmopressin, or replacement therapy (Table II). The majority of these patients were diagnosed with type 3 VWD. Only 5 control children (9%) required medical attention for epistaxis through visits to a healthcare professional. None of these children received any medical treatment. There was no effect of age ($P = .64$) or sex ($P = .35$) on PBQ epistaxis scores within patient and control groups.

Table II summarizes the features of epistaxis in both patients and control subjects. The percentage of patients with epistaxis frequency >5 episodes per year (67%) was significantly greater than that of control subjects (20%) ($P < .0001$). A total of 62% of patients had epistaxis lasting longer than 10 minutes (with 8 of these patients having episode[s] lasting longer than 1 hour), in contrast with only 9% of control children ($P < .0001$). The majority of both patients and control patients reported spontaneous onset of epistaxis ($P = .23$). A significantly greater percentage of patients bled from both nostrils, 77% vs 27% of control subjects ($P < .0001$). Seasonal correlation of epistaxis occurred less frequently in the patients compared with control children ($P = .022$).

A total of 44 of 107 patients (41%) were eligible for Katsanis ESS grading, having recurrent epistaxis defined as ≥5 nosebleeds per year. A total of 33 of these patients (75%) were graded as having severe epistaxis (score 7-10) and 10 patients (23%) as having mild epistaxis (score 0-6). There was no score documented for 1 patient, even though he was eligible for grading. Katsanis ESS–ineligible patients were patients who did not report a problem with epistaxis (n = 41) and 22 patients who had <5 nosebleeds per year. Only 11 of 163 control subjects (7%) had >5 nosebleeds per year and would have been eligible for Katsanis ESS grading.

Figure 3, A, shows the Katsanis ESS grading of 66 patients who reported a problem with epistaxis according to the PBQ. Information about diagnosis, number of patients, and median PBQ epistaxis score per Katsanis ESS grade (either ineligible, mild, or severe) are shown in Figure 3, B-D.

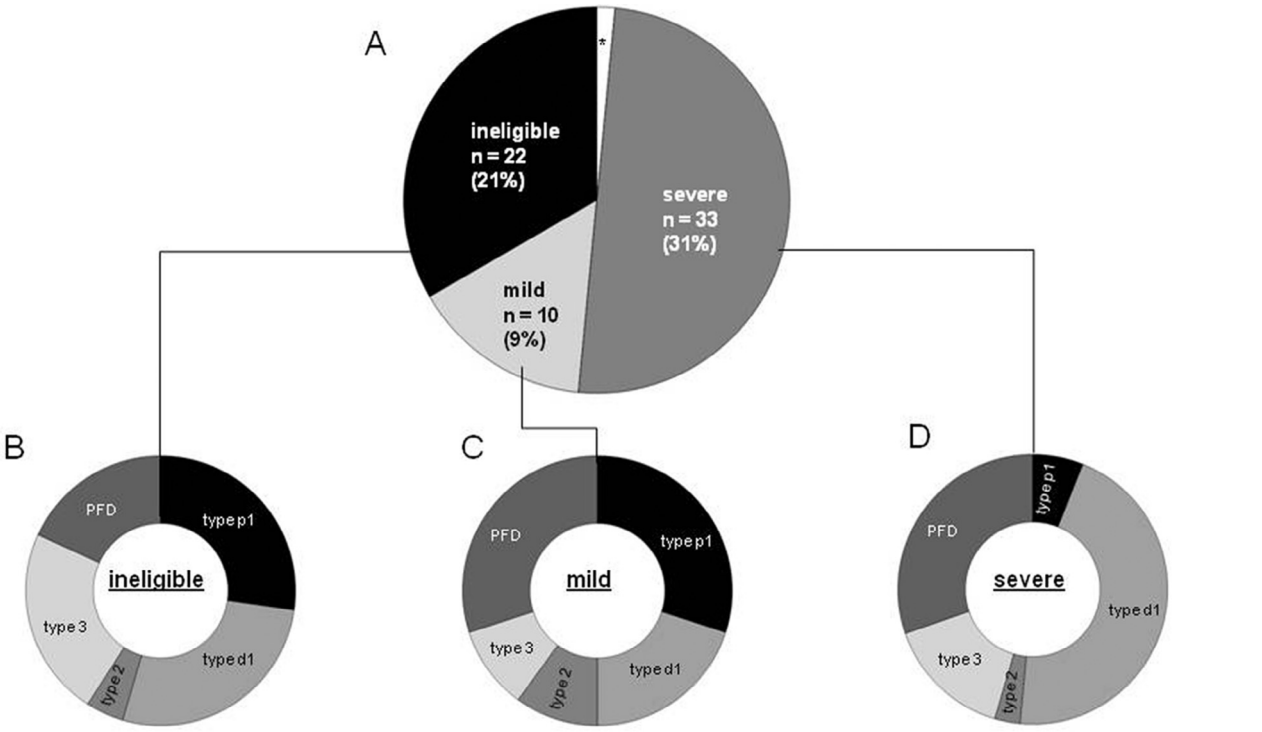
Of patients with definite type 1 VWD, 74% were eligible for grading via the Katsanis ESS compared with 46% of patients with possible type 1 VWD, 58% of patients with type 3 VWD, and 77% of patients with a PFD. A total of 91% of patients graded as severe epistaxis by the Katsanis ESS had clinically significant epistaxis according to the PBQ, whereas only 3 of 10 patients (30%) graded as mild epistaxis had clinically significant epistaxis.

Among the 22 Katsanis ESS–ineligible patients were 6 patients with definite type 1 VWD, 5 patients with type 3 VWD, and 4 patients with a PFD (Figure 3, B); 4 of these patients had a clinically significant PBQ epistaxis score. One of these Katsanis ESS–ineligible patients, who was diagnosed with type 3 VWD, received replacement therapy for an episode of epistaxis that lasted for several hours. Of the patients graded as having mild epistaxis, 3 of 10 had a clinically significant PBQ epistaxis score, as did 30 of 33 patients graded as having severe epistaxis. The 1 patient who did not have a documented Katsanis ESS score was diagnosed with type 3 VWD and received replacement therapy.

Discussion

Diagnosis of mucocutaneous bleeding disorders can be challenging, as affected children often present with bleeding symptoms, like epistaxis, which also occur in healthy children. The common bleeding symptom of epistaxis affects both sexes, can occur early in life (unlike menorrhagia), and does not require exposure to a hemostatic challenge (unlike dental extraction and postsurgical bleeding). There is an ongoing need for investigations that contribute to the understanding of the patterns of epistaxis in children with an underlying bleeding disorder as well as in normal children so that a screening tool can be developed to aid in determining whether a pediatric subject should or should not be referred for investigation of an underlying inherited or acquired bleeding disorder. To describe the severity and features of epistaxis in children with a known diagnosis of VWD or a PFD and healthy control children, responses to 2 organ-specific questionnaires, the PBQ epistaxis section and the Katsanis ESS, were reviewed. Epistaxis scores were calculated from both questionnaires, and in addition, specific features of epistaxis included in the PBQ epistaxis section were compared with results from control subjects to identify if any/which features could be discriminatory to identifying an underlying bleeding disorder.

Patients with definite type 1, type 2, and type 3 VWD as well as patients with a PFD had clinically more severe epistaxis compared with control subjects. This was reflected by significantly greater median PBQ epistaxis scores for each of these patient groups vs healthy control children. To date, not all information that is collected in the PBQ epistaxis section is used to calculate the bleeding score. We have analyzed the additional information about specific features of epistaxis, finding that most of these occur in a significantly greater proportion in patients with VWD or a PFD than in healthy children with epistaxis. Several of these features, such as longer duration of epistaxis, lack of seasonal correlation, and requirement for



Diagnosis	Number of patients	Median PBQ ES (range)
definite type 1 VWD	6	1 (0-2)
possible type 1 VWD	6	0 (0-0)
type 2 VWD	1	0
type 3 VWD	5	1 (0-4)
PFD	4	0.5 (0-4)

Diagnosis	Number of patients	Median PBQ ES (range)
definite type 1 VWD	2	1.5 (1-2)
possible type 1 VWD	3	1 (1-1)
type 2 VWD	1	2
type 3 VWD	1	4
PFD	3	1 (1-1)

Diagnosis	Number of patients	Median PBQ ES (range)
definite type 1 VWD	15	3 (1-4)
possible type 1 VWD	2	3 (2-4)
type 2 VWD	1	4
type 3 VWD	5	4 (4-4)
PFD	10	3 (1-4)

Figure 3. Comparison of Katsanis ESS and PBQ epistaxis score grading in patients with epistaxis. **A,** Katsanis ESS grading of 66 patients with epistaxis; % of total patient cohort (n = 107). Katsanis ESS grading according to patients' diagnosis (in pie chart) and PBQ epistaxis score (in table below the pie chart) are shown for patients with epistaxis who were graded as **B,** Katsanis ESS ineligible or **C,** having mild or **D,** severe epistaxis. *No Katsanis ESS score was documented for 1 eligible patient; type p1, possible type 1 VWD (referred to patients who had [1] a VWF:RCo of 0.05-0.50 U mL^{-1} on at least 2 occasions and a 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 but with a negative bleeding history according to SickKids (HSC) criteria or [2] abnormal VWF:RCo and VWF:Ag on at least 1 occasion with or without a bleeding history); type d1, definite type 1 VWD (referred to patients who had laboratory data that fit the aforementioned criteria and a positive bleeding history according to SickKids criteria).

medical intervention, were previously also found to be significantly associated with VWD in childhood.[5] Katsanis et al, however, reported that duration of bleeding alone might not reliably distinguish patients with mucocutaneous bleeding disorders, as some healthy children in their study had longer duration of nosebleeds than patients diagnosed with VWD.[10] This may be explained by the fact that only 36 children were included in their study, and only 2 were diagnosed with VWD. Regarding the requirement for medical intervention, Burton and Doree described that most healthy children with epistaxis do not seek medical attention and have not had any medical interventions.[14] In accordance with this observation, we found that only 9% of healthy children consulted a healthcare professional for epistaxis, in contrast with more than one-half of the patients with VWD or a PFD. Likely influenced by their underlying diagnosis, the majority of these patients received desmopressin, antifibrinolytics, or replacement therapy for treatment of their epistaxis, whereas none of the control subjects required any type of medical intervention.

We found 2 other features of epistaxis that occurred in a significantly greater proportion of patients compared with healthy children with epistaxis, specifically frequency of >5 episodes per year and epistaxis occurring from both nostrils either at the same time or separate time; these features have not been described previously in other studies. In contrast, in a retrospective chart review of 47 children with recurrent epistaxis who failed medical treatment (ie, emollient therapy) and subsequently had intraoperative cauterization of the septum, neither duration, frequency, nor location (unilateral/bilateral) of epistaxis were significant clinical predictors for a bleeding disorder, which was subsequently diagnosed in 5 of 47 patients.[15]

In 2012, Bidlingmaier et al compared the use of the PBQ with the ISTH-BAT for evaluation of a suspected bleeding disorder in a pediatric clinical setting.[16] Results of the ISTH-BAT correlated well with the PBQ scores, leading the authors to conclude that the ISTH-BAT could be useful in the clinic for deciding whether extended laboratory workup is reasonable. They encouraged the use of the ISTH-BAT in further studies for quantification of bleeding symptoms in different cohorts to improve comparability of studies. Because the ISTH-BAT epistaxis scoring key is identical to that of the PBQ, patients in our cohort would have had the exact same epistaxis scores if the ISTH-BAT was used instead of the PBQ. Thus, our results with the PBQ epistaxis score would be applicable to an ISTH-BAT epistaxis score. However, there are differences in the additional questions about specific features of epistaxis between the ISTH-BAT and PBQ. The ISTH-BAT epistaxis section does not include most of the questions about features that are in the PBQ, but it does ask about the age of first symptoms and approximate number of episodes not requiring medical attention, questions that are not in the PBQ; it would be interesting to investigate whether these 2 ISTH-BAT features of epistaxis differ between children with and without a bleeding disorder.

When the Katsanis ESS was used, one-half of the patients with epistaxis in our cohort were classified as having severe epistaxis. However, one-third of the patients with epistaxis were ineligible for the Katsanis ESS, having <5 nosebleeds per year. We observed that although patients with type 3 VWD had greater PBQ epistaxis scores than those with definite type 1 VWD or a PFD, 42% of these patients were ineligible for grading by the Katsanis ESS, compared with only 26% of the patients with definite type 1 VWD and 23% of the patients with a PFD. This indicates that the patients with definite type 1 VWD or a PFD were likely to have more frequent, but less clinically severe, epistaxis episodes that did not require therapy as compared with patients with type 3 VWD. This highlights an important limitation of the Katsanis ESS for general use because a particular minimum "threshold" of epistaxis frequency has to be reached before this scoring system can be applied, unlike the PBQ, which is able to assess any number of epistaxis episodes.

One of the benefits of using an organ-specific tool for children presenting with epistaxis is the relative shorter length of time it would take to answer the relevant questions, when compared with completion of the full PBQ or ISTH-BAT questionnaire. On average, <5 minutes is required to complete the Katsanis questionnaire or PBQ epistaxis section vs up to 20 minutes to complete either the PBQ or ISTH-BAT.[17]

This study is limited by the lack of Katsanis ESS data in 11 control subjects who met the eligibility criteria for grading. In addition, it is important to recognize that selection bias was an inevitable element in this study. To have a historical diagnosis of an inherited bleeding disorder, the majority of the children studied had presented initially with significant mucocutaneous bleeding symptoms that had led to their investigation and subsequent diagnosis. Although the interviewer had previous knowledge of the patients' diagnoses, they were not aware of their bleeding histories. Furthermore, an important limitation of this study is that children with a previous bleeding disorder diagnosis will generate a greater score due to the use of desmopressin/VWF concentrate, etc, for treatment compared with (undiagnosed) children who visit a physician (eg, family physicians, general pediatrician) for their bleeding symptoms for the first time. This should be kept in mind when interpreting the data presented in this manuscript.

In conclusion, results of this study show that the use of a standardized questionnaire to obtain an organ-specific bleeding history may be useful in the assessment of epistaxis severity in children with VWD or a PFD. Specific features of epistaxis may play an important role in discriminating between affected and healthy children. In this regard, an organ-specific questionnaire may help clinicians, particularly general pediatricians; family physicians; and ear, nose and throat surgeons in deciding which children presenting with epistaxis should undergo further investigation for a bleeding disorder. Such a questionnaire could incorporate features of epistaxis from both the PBQ and Katsanis questionnaires and a score but should not have the Katsanis frequency cut-off for patients' eligibility. Prospective evaluation of such a tool in previously undiagnosed children with epistaxis is required. ■

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A Epistaxis section of PBQ

Bleeding symptoms

Epistaxis	No ☐	If Yes, Trivial ☐	Significant ☐

AVERAGE PRESENTATION

Age of maximum severity	☐ 0 - 4 years ☐ 4 - 8 years ☐ 8 - 12 years ☐ 12 - 16 years ☐ 16 - 20 years	
Number episodes/year	☐ < 1 ☐ 1 - 5 ☐ 6 - 12 ☐ > 12	
Duration of average single episode	☐ < 1 minute ☐ 1-10 minutes ☐ > 10 minutes	
Spontaneous?	Yes ☐	No ☐
Both nostrils?	Yes ☐	No ☐
After drug ingestion (e.g.aspirin)	Yes ☐	No ☐
Seasonal correlation	Yes ☐	No ☐
	Specify:	
Cessation	☐ spontaneous ☐ after compression ☐ by medical intervention	

REPORT TREATMENT OF THE MOST SEVERE EPISODE

Required medical attention?	Yes ☐	No ☐
If yes, please specify:		
Consultation only	☐	
Packing	☐ # of times	
Cauterization	☐ # of times	
Antifibrinolytics	☐ # of times	
Desmopressin	☐ # of times	
Replacement therapy	☐ # of times	
Blood transfusion	☐ # of times	
Notes		

B PBQ epistaxis scoring key

Score / Symptom	-1	0	1	2	3	4
Epistaxis	-	No or trivial (≤5 per year)	>5 per year OR >10 minutes duration	Consultation only	Packing, cauterization or antifibrinolytics	Blood transfusion, replacement therapy or desmopressin

Figure 1. **A,** PBQ epistaxis section[5] and **B,** scoring key[5]; **C,** Katsanis ESS.[10] [1]Mild = 0-6; severe = 7-10. [2]Estimation of average blood loss per episode, based on fractions or multiples of teaspoons, tablespoons, or cups. [3]Proportion of child's life that nosebleeds have been recurrent (≥5 per year). (*Continues*)

C Katsanis ESS

Component	Score[1]
Frequency	
5-15/yr	0
16-25/yr	1
> 25/yr	2
Duration	
< 5 min	0
5-10 min	1
> 10 min	2
Amount[2]	
< 15 ml	0
15-30 ml	1
> 30 ml	2
Epistaxis history/age[3]	
< 33%	0
33%-67%	1
> 67%	2
Site	
Unilateral	0
Bilateral	2

Figure 1. Continued.