

RODEGHIERO, F., TOSETTO, A., ABSHIRE, T., ARNOLD, D. M., COLLER, B., JAMES, P., NEUNERT, C., LILLICRAP, D. AND ON BEHALF OF THE ISTH/SSC JOINT VWF AND PERINATAL/PEDIATRIC HEMOSTASIS SUBCOMMITTEES WORKING GROUP (2010), ISTH/SSC BLEEDING ASSESSMENT TOOL: A STANDARDIZED QUESTIONNAIRE AND A PROPOSAL FOR A NEW BLEEDING SCORE FOR INHERITED BLEEDING DISORDERS. JOURNAL OF THROMBOSIS AND HAEMOSTASIS, 8: 2063–2065.

SUPPLEMENTARY MATERIAL TO THE OFFICIAL COMMUNICATION OF THE SSC (LAST REVISION: 19 JULY 2011)

Background

The clinical appreciation of the presence and severity of bleeding symptoms is a fundamental step in the evaluation of patients referred for a possible bleeding disorder. In an attempt to improve the collection and reproducibility of the bleeding history, several Bleeding Assessment Tools (BAT) have been proposed and used. Currently available BAT have some limitations, particularly regarding the lack of pediatric-specific symptoms in some of them and the predominance of the severity of bleeding symptoms over other potentially clinically important features, such as the frequency of symptoms.

To overcome the above-mentioned limitations and to promote the standardization of the available BATs, a Working Group was established within the framework of the ISTH/SSC Subcommittees on VWF and on Perinatal/Pediatric Hemostasis (ISTH/SSC-BAT) during the 53rd SSC Annual Meeting held in Geneva in 2007. Members of the group first met in Toronto on January 2008 and then regularly at each subsequent SSC meeting. This paper presents a structured questionnaire and its clinical use agreed on by the ISTH/SSC-BAT together with a proposal for a new BS system to undergo validity and reliability testing in future studies. This new BAT is intended for inherited bleeding disorders in children and adults. The questionnaire should be collected by a physician or another adequately trained health-

professional. Only symptoms and related treatments, if any, before and/or at diagnosis should be reported. Refer to the full text for additional instructions.

Minimal criteria defining a significant bleeding

For each specific bleeding symptom, the ISTH/SSC joint working group proposed minimal criteria in order to classify a symptom as significant and thus receive a score of 1 or more (see also Table 1):

1. Epistaxis: Any nosebleed, especially occurring after puberty, that causes patient concern (e.g., interference or distress with daily or social activities) is considered significant. In general, epistaxis should not be considered significant when it lasts less than 10 minutes, has a frequency of < 5 episodes/year, has a seasonal occurrence, or is associated with infections of the upper respiratory tract or other identifiable cause (e.g., dusty dry air).
2. Cutaneous bleeding: Bruises are considered significant when 5 or more (> 1cm) in exposed areas; petechiae when adequately described by the patient or relatives; or hematomas when occurring without trauma.
3. Minor cutaneous wound: Any bleeding episode caused by superficial cuts (e.g., by shaving razor, knife, or scissors) or that requires frequent bandage changes is considered significant. Insignificant bleeding from wounds includes those of duration < 10 minutes and lesions that usually require stitches in normal subjects (e.g., under the chin). Symptoms should also be manifest on more than one occasion to be considered significant.
4. Oral cavity bleeding: Gum bleeding should be considered significant when it causes frankly bloody sputum and lasts for 10 minutes or longer on more than one occasion. Tooth eruption or spontaneous tooth loss bleeding should be considered significant when it requires assistance or supervision by a physician, or lasts at least 10 minutes (bleeding associated with tooth extraction is considered separately). Bleeding occurring after bites to lips, cheek, and tongue should be considered significant when it lasts at least 10 minutes or causes a swollen tongue or mouth.
5. Hematemesis, melena, and hematochezia: Any gastrointestinal bleeding that is not explained by the presence of a specific disease should be considered significant.

6. Hematuria: Only macroscopic hematuria (from red to pale-pink urine) that is not explained by the presence of a specific urologic disease should be considered significant.
7. Tooth extraction: Any bleeding occurring after leaving the dentist's office and requiring a new, unscheduled visit or prolonged bleeding at the dentist's office causing a delay in the procedure or discharge should be considered significant.
8. Surgical bleeding: Any bleeding judged by the surgeon to be abnormally prolonged, that causes a delay in discharge, or requires some supportive treatment is considered significant.
9. Menorrhagia: Any bleeding that interferes with daily activities such as work, housework, exercise or social activities during most menstrual periods should be considered significant. Criteria for significant bleeding may include any of the following: changing pads more frequently than every 2 hours; menstrual bleeding lasting 7 or more days; and the presence of clots > 1 cm combined with a history of flooding. If a patient has previously made a record of her menstrual loss using a pictorial blood loss assessment chart (PBAC), a PBAC score higher than 100 also qualifies for a score of 1.
10. Post-partum bleeding. Vaginal bleeding or uterine discharge (lochia) that lasts for more than 6 weeks. Any bleeding of lesser duration that is judged by the obstetrician as abnormally heavy or prolonged, that causes a delay in discharge, requires some supportive treatment, requires changing pads or tampons more frequently than every 2 hours, or causes progressive anemia is also considered significant
11. Muscle hematomas or hemarthrosis. Any spontaneous joint / muscle bleeding (not related to traumatic injuries) is considered significant.
12. CNS bleeding. Any subdural or intracerebral hemorrhage requiring diagnostic or therapeutic intervention is scored 3 or 4, respectively.
13. Other bleeding symptoms. When these bleeding symptoms occur during infancy, they are scored 1 or more. Their presence when reported by either the patient or a family member should always prompt detailed laboratory investigation.

Only symptoms and treatment BEFORE and AT diagnosis should be considered

1. Epistaxis

- 1.1 Have you ever had spontaneous epistaxis? Yes No or trivial (skip to 2)
- 1.2 Have the symptom ever required medical attention ? Yes No (resolve spontaneously; skip to 1.6)
- 1.3 If answer to 1.2 is yes, please specify
- Consultation only
 - Cauterization
 - Packing
 - Antifibrinolytics
 - Iron therapy
 - Treatment with desmopressin
 - Treatment with plasma
 - Treatment with platelet concentrate
 - Treatment with factor concentrates
 - Blood (RBC) transfusion
- 1.4 How many times in your life did you receive any of the above treatments (# 1.3)?
- 1 - 2
 - 3 to 5
 - 6 to 10
 - more than 10
- 1.5 At what age did you first have symptoms?
- Before 1 year
 - Between 1-5 years of age
 - Between 6-12 years of age
 - Between 13-25 years of age
 - After 25 years of age
- 1.6 Approximate number of episodes NOT requiring medical attention
- less than 1 per year
 - 1 per year
 - 2-5 every year
 - 1-3 every month
 - 1 every week
- 1.7 Duration of average single episode (min.) NOT requiring medical attention
- 1 minute or less
 - 1 - 10 minutes
 - more than 10 minutes

2. Cutaneous bleeding (Bruising, ecchymoses, purpura, subcutaneous hematomas)

- 2.1 Have you ever had any of the above cutaneous bleeding? Yes No or trivial skip to 3
- 2.2 Have the symptom ever required medical attention? Yes No skip to 2.6
- 2.3 If answer to 2.2 is yes, please specify
- Consultation only
 - Treatment with desmopressin
 - Treatment with plasma
 - Treatment with platelet concentrate
 - Treatment with factor concentrates
 - Blood (RBC) transfusion
- 2.4 How many times in your life did you receive any of the above treatments (# 2.3)?
- 1 - 2
 - 3 to 5
 - 6 to 10
 - more than 10
- 2.5 At what age did you first have symptoms?
- Before 1 year
 - Between 1-5 years of age
 - Between 6-12 years of age
 - Between 13-25 years of age
 - After 25 years of age
- 2.6 Approximate number of episodes NOT requiring medical attention
- less than 1 per year
 - 1 per year
 - 1-5 every six month
 - 1-3 every month
 - 1 every week
- 2.7 Type of bleeding
- Petechiae
 - Bruises
 - Hematomas
- 2.8 Location
- Exposed sites
 - Unexposed sites
 - Both
- 2.9 Common size
- ≤ 1 cm
 - >1 cm
 - Extensive (palm sized or larger)
- 2.10 How many bruises >1 cm in exposed areas in the most severe manifestation?
- ≤ 5
 - > 5
- 2.11 Location of petechiae
- Limited to lower limbs
 - Diffuse

3. Bleeding from minor wounds (not requiring stitches in the average patient)

- 3.1 Have you ever had prolonged bleeding from minor wounds? Yes No or trivial skip to 4
- 3.2 Have the symptom ever required medical attention ? Yes No skip to 3.6
- 3.3 If answer to 3.2 is yes, please specify
- Consultation only
 - Surgical hemostasis
 - Treatment with desmopressin
 - Treatment with plasma
 - Treatment with platelet concentrate
 - Treatment with factor concentrates
 - Blood (RBC) transfusion
- 3.4 How many times in your life did you received any of the above treatments (# 3.3)?
- 1 - 2
 - 3 to 5
 - 6 to 10
 - more than 10
- 3.5 At what age did you first have symptoms?
- Before 1 year
 - Between 1-5 years of age
 - Between 6-12 years of age
 - Between 13-25 years of age
 - After 25 years of age
- 3.6 Approximate number of episodes NOT requiring medical attention
- less than 1 per year
 - 1 per year
 - 2-5 every year
 - 1-3 every month
 - 1 every week
- 3.7 Duration of average single episode (min.)
- 1 to 10 minutes
 - more than 10 minutes

4. Hematuria

4.1 Have you ever had hematuria ? Yes No skip to 5

4.2 If answer to 4.1 is yes, please specify

Presence of associated urologic disease

Yes (skip to 5)

No

Specify:

- Stones
- Infection
- Kidney/ bladder disease

Please answer the following questions only for SPONTANEOUS symptoms (answer No to 4.1)

4.3 Have the symptom ever required medical attention ? Yes No skip to 4.7

4.4 If answer to 4.3 is yes, please specify

Consultation only

- Surgery
- Iron therapy

Treatment with desmopressin

- Treatment with plasma
- Treatment with platelet concentrate
- Treatment with factor concentrates

Blood (RBC) transfusion

4.5 How many times in your life did you received any of the above treatments (# 4.4)?

- 1 - 2
- 3 to 5
- 6 to 10
- more than 10

4.6 At what age did you first have symptoms?

- Before 1 year
- Between 1-5 years of age
- Between 6-12 years of age
- Between 13-25 years of age
- After 25 years of age

4.7 Approximate number of episodes NOT requiring medical attention

- less than 1 per year
- 1 per year
- 1-5 every six month
- 1-3 every month
- 1 every week

5. Gastrointestinal bleeding (Hematemesis, Melena, Hematochezia)

5.1 Have you ever had gastrointestinal bleeding ? Yes No skip to 6

5.2 If answer to 5.1 is yes, please specify

Type of bleeding

- Hematemesis
- Melena
- Hematochezia

Presence of associated GI disease

Yes No

Specify:

- Ulcer
- Portal hypertension
- Angiodysplasia

Please answer to the following questions only for SPONTANEOUS symptoms

5.3 Have the symptom ever required medical attention ? Yes No skip to 5.7

5.4 If answer to 5.3 is yes, please specify

- Consultation only
- Surgical haemostasis
- Antifibrinolytics
- Treatment with desmopressin
- Treatment with plasma
- Treatment with platelet concentrate
- Treatment with factor concentrates
- Blood (RBC) transfusion

5.5 How many times in your life did you received any of the above treatments (# 5.4)?

- 1 - 2
- 3 to 5
- 6 to 10
- more than 10

5.6 At what age did you first have symptoms?

- Before 1 year
- Between 1-5 years of age
- Between 6-12 years of age
- Between 13-25 years of age
- After 25 years of age

5.7 Approximate number of episodes NOT requiring medical attention

- less than 1 per year
- 1 per year
- 1-5 every six month
- 1-3 every month
- 1 every week

6. Oral cavity bleeding (Tooth eruption, spontaneous or after brushing/flossing, gum bleeding, bleeding after bites to lip & tongue)

- 6.1 Have you ever had oral cavity bleeding ? Yes No or trivial skip to 7
- 6.2 Have the symptom ever required medical attention ? Yes No skip to 6.6
- 6.3 If answer to 6.2 is yes, please specify
- Consultation only
 - Surgical hemostasis (dental packing, suture, cauterization)
 - Antifibrinolytics
 - Treatment with desmopressin
 - Treatment with plasma
 - Treatment with platelet concentrate
 - Treatment with factor concentrates
 - Blood (RBC) transfusion
- 6.4 How many times in your life did you received any of the above treatments (# 6.3)?
- 1 - 2
 - 3 to 5
 - 6 to 10
 - more than 10
- 6.5 At what age did you first have symptoms?
- Before 1 year
 - Between 1-5 years of age
 - Between 6-12 years of age
 - Between 13-25 years of age
 - After 25 years of age
- 6.6 Approximate number of episodes NOT requiring medical attention
- less than 1 per year
 - 1 per year
 - 1-5 every six month
 - 1-3 every month
 - 1 every week
- 6.7 Duration of average single episode (min.)
- 1 to 10 minutes
 - more than 10 minutes

7. Bleeding after Tooth/ Teeth extraction

7.1 Have you ever had bleeding after tooth (teeth) extraction ? Yes No

7.2 Please specify number of extractions If no extractions, skip to section 8

Please fill in one of the following forms for **each** tooth extraction

Age at extraction	<input type="text"/> <input type="text"/>	Type of extraction	<input type="checkbox"/> Deciduous <input type="checkbox"/> Permanent <input type="checkbox"/> Molar
Actions taken to prevent bleeding	<input type="checkbox"/> None <input type="checkbox"/> Antifibrinolytics <input type="checkbox"/> Desmopressin <input type="checkbox"/> Plasma or clotting factor concentrates <input type="checkbox"/> Platelet infusion		
Bleeding after extraction?	Yes <input type="checkbox"/>	No <input type="checkbox"/>	
Actions taken to control bleeding	<input type="checkbox"/> None <input type="checkbox"/> Resuturing <input type="checkbox"/> Packing <input type="checkbox"/> Antifibrinolytics <input type="checkbox"/> Desmopressin <input type="checkbox"/> Plasma or clotting factor concentrates <input type="checkbox"/> Platelet infusion <input type="checkbox"/> Blood (RBC) transfusion		

8. Bleeding after Surgery or Major Trauma

8.1 Have you ever had bleeding after surgery or major trauma ? Yes No

8.2 Please specify number of surgeries/ major trauma If no Surgery or Trauma skip to section 9

Please fill in one of the following forms for **each** surgery or major trauma episode

Age at intervention/trauma	<input type="text"/> <input type="text"/>	Type of surgery	<input type="checkbox"/> Major-abdominal
			<input type="checkbox"/> Major-thoracic
		<input type="checkbox"/> Tonsillectomy/Adenoids	<input type="checkbox"/> Major-gynecology
		<input type="checkbox"/> Pharynx/Nose	<input type="checkbox"/> Other
Actions taken to prevent bleeding		<input type="checkbox"/> None	
		<input type="checkbox"/> Antifibrinolytics	
		<input type="checkbox"/> Desmopressin	
		<input type="checkbox"/> Plasma or clotting factor concentrates	
		<input type="checkbox"/> Platelet infusion	
Bleeding after intervention?		Yes <input type="checkbox"/>	No <input type="checkbox"/>
Actions taken to control bleeding		<input type="checkbox"/> None	
		<input type="checkbox"/> Surgical hemostasis	
		<input type="checkbox"/> Antifibrinolytics	
		<input type="checkbox"/> Desmopressin	
		<input type="checkbox"/> Plasma or clotting factor concentrates	
		<input type="checkbox"/> Platelet infusion	
		<input type="checkbox"/> Blood (RBC) transfusion	

9. Menorrhagia

- 9.1 Have you ever had very heavy menstrual bleeding (menorrhagia)? Yes No or trivial skip to 10
- If answer to 9.1 is yes, please specify
- Changing pads/tampons more frequently than every 2 hours
 - Bleeding more than 7 days
 - Clot and flooding
- Impairment of daily activities (work, housework, exercise, social activities): Never or rarely Most menses
- 9.2 Have the symptom ever required medical attention ? Yes No skip to 9.6
- 9.3 If answer to 9.2 is yes, please specify
- a Consultation only
 - b Pictorial Bleeding Score _____ Assessment
 - c Antifibrinolytic therapy
 - d Iron therapy
 - e Hormonal therapy
 - f Combined antifibrinolytics & Hormonal therapy
 - g Hysterectomy / endometrial ablation / D & C
 - h Treatment with desmopressin
 Treatment with plasma
 Treatment with platelet concentrate
 Treatment with factor concentrates
 - i Blood (RBC) transfusion
 - l Hospital admission and emergency treatment
- 9.4 How many times in your life did you received any of the above treatments (# 9.3 a-l)?
- 1 - 2
 - 3 to 5
 - 6 to 10
 - more than 10
- 9.5 At what age did you first have symptoms?
- At menarche
 - Between 14-25 years of age
 - After 25 years of age
- 9.6 Have you had time off work/school for menorrhagia?
- < twice a year
 - > twice a year
- 9.7 Duration of menorrhagia
- Since menarche
 - > 12 months
 - < 12 months
- 9.8 Have you had acute menorrhagia requiring emergency treatment/hospital admission Yes No
- How many times: _____

10. Post-partum hemorrhage

- 10.1 Number of successful pregnancies (live births)
- 10.2 Have you ever had post-partum haemorrhage? Yes No or trivial skip to 11
- 10.3 Did it occur In the first 24 hours after delivery (Primary)
 Between 24 hours and 6 weeks postpartum (Secondary)
 Both Primary and Secondary
- 10.4 How long did vaginal discharge (lochia) last? < 6 weeks
 > 6 weeks
- 10.5 Did it require changing pads/tampons more frequently than every 2 hours? Yes No
- 10.6 Did this bleeding cause delay of hospital discharge/ readmission to hospital? Yes No
- 10.7 Have the symptom ever required medical treatment? Yes No
- 10.8 If answer to 10.7 is yes, please specify Consultation only /oxytocin i.v. infusion
 Additional uterotonic medications
 Iron therapy
 Antifibrinolytic therapy
 Treatment with desmopressin
 Treatment with plasma
 Treatment with platelet concentrate
 Treatment with factor concentrates
 Blood (RBC) transfusion
 Any procedure requiring examination under anaesthesia
 Uterine balloon/package to tamponade the uterus
 Any procedure requiring critical care or surgical intervention (includes: hysterectomy, internal iliac artery ligation, uterine artery embolization, uterine brace sutures)
- 10.9 Number of deliveries that required any of the above treatments (# 10.8)?

11. Muscle hematomas (spontaneous)

- 11.1 Have you ever had muscle hematomas or hemarthrosis ? Yes No or trivial skip to 12
- If yes, was it spontaneous or after trauma? Yes, spontaneous No, trauma-related
- 11.2 Have the symptom ever required medical attention ? Yes No skip to 11.6
- 11.3 If answer to 11.2 is yes, please specify
- Consultation only
 - Surgical draining
 - Treatment with desmopressin
 - Treatment with plasma
 - Treatment with platelet concentrate
 - Treatment with factor concentrates
 - Blood transfusion
- 11.4 How many times in your life did you receive any of the above treatments (# 11.3)?
- 1 - 2
 - 3 to 5
 - 6 to 10
 - more than 10
- 11.5 At what age did you first have symptoms?
- Before 1 year
 - Between 1-5 years of age
 - Between 6-12 years of age
 - Between 13-25 years of age
 - After 25 years of age
- 11.6 Approximate number of episodes NOT requiring medical attention
- less than 1 per year
 - 1 per year
 - 1-5 every six month
 - 1-3 every month
 - 1 every week

12. Hemarthrosis

- 12.1 Have you ever had muscle hematomas or hemarthrosis ? Yes No or trivial skip to 13
- If yes, was it spontaneous or after trauma? Yes, spontaneous No, trauma-related
- 12.2 Have the symptom ever required medical attention ? Yes No skip to 12.6
- 12.3 If answer to 12.2 is yes, please specify
- Consultation only
 - Surgical draining
 - Treatment with desmopressin
 - Treatment with plasma
 - Treatment with platelet concentrate
 - Treatment with factor concentrates
 - Blood transfusion
- 12.4 How many times in your life did you receive any of the above treatments (# 11.3)?
- 1 - 2
 - 3 to 5
 - 6 to 10
 - more than 10
- 12.5 At what age did you first have symptoms?
- Before 1 year
 - Between 1-5 years of age
 - Between 6-12 years of age
 - Between 13-25 years of age
 - After 25 years of age
- 12.6 Approximate number of episodes NOT requiring medical attention
- less than 1 per year
 - 1 per year
 - 1-5 every six month
 - 1-3 every month
 - 1 every week

13. CNS bleeding (spontaneous)

- 13.1 Have you ever had cranial or spinal bleeding? Yes No or trivial skip to 14
- If yes, was it spontaneous or after trauma? Yes, spontaneous No, trauma-related
- 13.2 If answer to 13.1 is yes, please specify
- Type of bleeding Subdural
 Intracerebral
 Subarachnoid
- Was the diagnosis made by CT scan
 MNR
 Angiography
- 13.3 Type of treatment
- Consultation
- Surgical draining
- Treatment with plasma, platelet or factor concentrates
- 13.4 At what age did you have CNS bleeding?
- Before 1 year
 Between 1-5 years of age
 Between 6-12 years of age
 Between 13-25 years of age
 After 25 years of age

14 Other bleedings

14.1 Have you ever had one of the following?

Excessive umbilical stump bleeding Yes No

Cephalohematoma Yes No

Bleeding at circumcision Yes No

Venipuncture bleeding Yes No

Suction Bleeding Yes No

Ovulation bleeding(in women) Yes No

14.2 Have one of these symptoms ever required medical attention? Yes No

14.3 If answer to 12.2 is yes, please specify Consultation only

Antifibrinolytics

Surgery

Treatment with desmopressin

Treatment with plasma

Treatment with platelet concentrate

Treatment with factor concentrates

Blood (RBC) transfusion

14.4 How many times in your life did you receive any of the above treatments (# 12.3) for this symptom?

1 - 2

3 to 5

6 to 10

more than 10

Table 1. Bleeding score

SYMPTOMS (up to the time of diagnosis)	SCORE				
	0 ^s	1 ^s	2	3	4
Epistaxis	No/trivial	- > 5/year or - more than 10 minutes	Consultation only*	Packing or cauterization or antifibrinolytic	Blood transfusion or replacement therapy (use of hemostatic blood components and rFVIIa) or desmopressin
Cutaneous	No/trivial	For bruises 5 or more (> 1cm) in exposed areas	Consultation only*	Extensive	Spontaneous hematoma requiring blood transfusion
Bleeding from minor wounds	No/trivial	- > 5/year or - more than 10 minutes	Consultation only*	Surgical hemostasis	Blood transfusion, replacement therapy, or desmopressin
Oral cavity	No/trivial	Present	Consultation only*	Surgical hemostasis or antifibrinolytic	Blood transfusion, replacement therapy or desmopressin
GI bleeding	No/trivial	Present (not associated with ulcer, portal hypertension, hemorrhoids, angiodysplasia)	Consultation only*	Surgical hemostasis, antifibrinolytic	Blood transfusion, replacement therapy or desmopressin

Hematuria	No/trivial	Present (macroscopic)	Consultation only*	Surgical hemostasis, iron therapy	Blood transfusion, replacement therapy or desmopressin
Tooth extraction	No/trivial or none done	Reported in $\leq 25\%$ of all procedures, no intervention**	Reported in $>25\%$ of all procedures, no intervention**	Resuturing or packing	Blood transfusion, replacement therapy or desmopressin
Surgery	No/trivial or none done	Reported in $\leq 25\%$ of all procedures, no intervention**	Reported in $>25\%$ of all procedures, no intervention**	Surgical hemostasis or antifibrinolytic	Blood transfusion, replacement therapy or desmopressin
Menorrhagia	No/trivial	Consultation only* or - Changing pads more frequently than every 2 hours or - Clot and flooding or - PBAC score $>100^{\#}$	- Time off work/school $> 2/\text{year}$ or - Requiring antifibrinolytics or hormonal or iron therapy	- Requiring combined treatment with antifibrinolytics and hormonal therapy or - Present since menarche and > 12 months	- Acute menorrhagia requiring hospital admission and emergency treatment or - Requiring blood transfusion, Replacement therapy, Desmopressin, or - Requiring dilatation & curettage or endometrial ablation or hysterectomy)
Post-partum hemorrhage	No/trivial or no deliveries	Consultation only* or - Use of syntocin or - Lochia > 6 weeks	- Iron therapy or - Antifibrinolytics	- Requiring blood transfusion, replacement therapy, desmopressin or - Requiring examination under anaesthesia and/or the use of uterin balloon/package to tamponade the uterus	- Any procedure requiring critical care or surgical intervention (e.g. hysterectomy, internal iliac artery legation, uterine artery embolization, uterine brace sutures)
Muscle hematomas	Never	Post trauma, no therapy	Spontaneous, no therapy	Spontaneous or traumatic, requiring desmopressin or replacement therapy	Spontaneous or traumatic, requiring surgical intervention or blood transfusion

Hemarthrosis	Never	Post trauma, no therapy	Spontaneous, no therapy	Spontaneous or traumatic, requiring desmopressin or replacement therapy	Spontaneous or traumatic, requiring surgical intervention or blood transfusion
CNS bleeding	Never	-	-	Subdural, any intervention	Intracerebral, any intervention
Other bleedings [^]	No/trivial	Present	Consultation only*	Surgical hemostasis, antifibrinolytics	Blood transfusion or replacement therapy or desmopressin

In addition to the guidance offered by the table, it is mandatory to refer to the text for more detailed instructions.

[§] Distinction between 0 and 1 is of critical importance. Score 1 means that the symptom is judged as present in the patient's history by the interviewer but does not qualify for a score 2 or more

* Consultation only: the patient sought medical evaluation and was either referred to a specialist or offered detailed laboratory investigation

** Example: 1 extraction/surgery resulting in bleeding (100%): the score to be assigned is 2; 2 extractions/surgeries, 1 resulting in bleeding (50%): the score to be assigned is 2; 3 extractions/surgeries, 1 resulting in bleeding (33%): the score to be assigned is 2; 4 extractions/surgeries, 1 resulting in bleeding (25%): the score to be assigned is 1

[#] If already available at the time of collection

[^] Include: umbilical stump bleeding, cephalohematoma, cheek hematoma caused by sucking during breast/bottle feeding, conjunctival hemorrhage or excessive bleeding following circumcision or venipuncture. Their presence in infancy requires detailed investigation independently from the overall score

Acknowledgments

We wish to acknowledge the collaboration of the other members of the ISTH/SSC Joint VWF and Perinatal/Pediatric Hemostasis Subcommittees Working Group: Christoph Bidlingmaier (Germany), Victor Blanchette (Canada), George Buchanan (USA), Jorge DiPaola (USA), Gili Kenet (Israel), Robert Montgomery (USA), James Riddel (USA), Margaret Rand (Canada), and Nicole Schlegel (France).

We are also most grateful to the members of the Menorrhagia Working Group of the Women's Issues SSC/ISTH Subcommittee: Rezan A. Kadir (UK), Peter Kouides (USA), Christine Lee (UK), Flora Peyvandi (Italy), Claire Philipp (USA), and Rochelle Winikoff (Canada) for their valuable contributions in the formulation of the questionnaire and BS, for menorrhagia and post-partum bleeding.

In addition, we wish to thank the personal contribution of Andra H. James (USA) and Sabine Eichinger (Austria).