

BLEEDING SCORE AND BLEEDING
QUESTIONNAIRE FOR THE DIAGNOSIS OF
TYPE 1 VON WILLEBRAND DISEASE

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BLEEDING SCORE AND BLEEDING QUESTIONNAIRE FOR THE DIAGNOSIS OF TYPE 1 VON WILLEBRAND DISEASE

PRESENTATION

A physician administered questionnaire for history taking and bleeding score assignment in patients presenting with bleeding symptoms. Minimal clinical criteria for the identification of subjects demanding further investigations for type 1 VWD can be obtained with the use of this questionnaire and the derived quantitative assessment.

See Appendix for the criteria to be used to compute the bleeding score.

Based on:

Rodeghiero F, Castaman G, Toso A, Battlè J, Baudo F, Cappelletti A, Casana P, De Bosch N, Eikenboom JCJ, Federici AB, Lethagen S, Linari S, Srivastava A: *The discriminant power of bleeding history for the diagnosis of von willebrand disease type 1: an international, multicenter study*. JTH, 2005

The questionnaire has been validated evaluating hemorrhagic symptoms in obligatory carriers of type 1 VWD and comparing them with those observed in age and sex-matched normal controls. A semiquantitative bleeding score was subsequently computed from the information collected from the questionnaire.

Limitations of the questionnaire are discussed in the paper.

For further information or comments please contact:

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QUESTIONNAIRE

Guidelines for history taking

Bleeding history is an essential component in the diagnosis of von Willebrand disease. To be valuable, history should not be a simple list of symptoms as spontaneously described by the patient or his/her relatives. It should be the result of a careful medical interview conducted by an expert physician posing critical questions to the patients.

Occurrence, frequency, severity and other inherent characteristics of every bleeding episode should be fully investigated. However, absence of bleeding in circumstances in which it could be expected is as well as important as its presence for the purpose of establishing a bleeding diathesis. It is essential that every symptom or its absence should be firmly established and thoroughly described. Thus, the proposed questionnaire represents only a simple guide, aiming at standardizing the art of history taking. It is not intended as a mean to exonerate the investigator to exert his/her own criticism in interpreting the patient's description by simply filling in the questionnaire based on patient's rough answers. The investigator should also appreciate the perception of the symptoms by the patient.

It is clearly impossible to report in detail every symptom, e.g. every episode of epistaxis or menorrhagia. Thus, necessarily the investigator should try to offer the most accurate and significant overall picture by describing the average episode and its frequency.

A similar approach is required to collect the history from control subjects to be included in some investigation in this area.

It is important to distinguish between real symptoms in a clinical sense and episodes of trivial importance simply reported by overzealous subjects. To this purpose and in order to assure standardisation, we offer a descriptive threshold (cut-off) below which a specific bleeding episode does not reach the level of a "symptom" and should not be reported in the questionnaire but only marked as "trivial" in the appropriate box.

Epistaxis: Any nosebleed, especially if not occurring during pre-puberal age only, which could not be managed by the patient him/herself OR longer than 5 minutes OR requiring medical attention. Very frequent and disturbing bleedings (at least one every week) could be recorded even if not meeting the above criteria in the "Note" space of the appropriate box of the questionnaire.

Bruising/ Hematoma: Any spontaneous bruise/hematoma larger than 3 centimetres or considered disproportionate to trauma by the investigator

Petechiae: No further requirements

Minor cutaneous wound: Any prolonged bleeding, longer than 5 minutes, caused by superficial cuts (e.g. by razor, knife or scissors)

Gum bleeding: Any spontaneous bleeding lasting for a minute or longer causing frankly bloody sputum or any profuse bleeding after tooth brushing

Tooth eruption: Any bleeding requiring assistance or supervision by a physician

Bites to lips, cheek and tongue: Any bleeding longer than 5 minutes or causing a swollen tongue or mouth

Hematemesis, melena and hematochezia: No further requirement

Tooth extraction: Any bleeding occurring after leaving the dentist's office or a prolonged bleeding at the dentist's office causing a delay in the procedure

Surgical bleeding: Any bleeding stated as abnormally prolonged by the surgeon or causing a delay in discharge or requiring some supportive treatment

Menorrhagia: See the questionnaire

Post-partum hemorrhage: As for surgery

For every symptom try to fill in the questionnaire summarizing it on the basis of the average presentation that is the most frequent one.

Please note that the present questionnaire pertains to subjects investigated before any definite diagnosis is made. Upon request, the second part of the questionnaire is available to collect bleeding symptoms in established VWD patients.

Epistaxis	No <input type="checkbox"/>	Trivial <input type="checkbox"/>	N.E. <input type="checkbox"/>	Yes <input type="checkbox"/>
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If yes, please fill in the following boxes

AVERAGE PRESENTATION	
Number episodes/year	<input type="checkbox"/> less than 1 <input type="checkbox"/> 1 - 5 <input type="checkbox"/> 6 - 12 <input type="checkbox"/> more than 12
Duration of average single episode (min.)	<input type="checkbox"/> one minute or less <input type="checkbox"/> one to ten minutes <input type="checkbox"/> more than ten minutes
Spontaneous ?	Yes <input type="checkbox"/> No <input type="checkbox"/>
Both nostrils ?	Yes <input type="checkbox"/> No <input type="checkbox"/>
After drug ingestion (e.g. aspirin)	Yes <input type="checkbox"/> No <input type="checkbox"/>
Seasonal correlation	Yes <input type="checkbox"/> No <input type="checkbox"/>
Cessation	<input type="checkbox"/> spontaneous <input type="checkbox"/> after short compression <input type="checkbox"/> by medical intervention
Age of maximum severity	<input type="checkbox"/> less than 14 years <input type="checkbox"/> 14 to 45 years <input type="checkbox"/> more than 45 years
Ever required medical attention ?	Yes <input type="checkbox"/> No <input type="checkbox"/>
if yes, please specify	
Consultation only	<input type="checkbox"/>
Cauterization	<input type="checkbox"/>
Packing	<input type="checkbox"/>
Blood transfusion	<input type="checkbox"/>
Notes

MOST SEVERE PRESENTATION

Duration of episode (min.)

- one minute or less
- one to ten minutes
- more than ten minutes

Spontaneous ?

Yes No

Both nostrils ?

Yes No

After drug ingestion
(e.g. aspirin)

Yes No

Cessation

- spontaneous
- after short compression
- by medical intervention

Ever required medical
attention ?

Yes No

if yes, please specify

Consultation only

Cauterization

Packing

Blood transfusion

Notes

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.....
.....

Cutaneous symptomsNo Trivial N.E. Yes

If yes, please fill in the following boxes

AVERAGE PRESENTATION

Number episodes/year

Type

- Petechiae
 Bruises
 Hematomas

Location of bruises (if any)

- Exposed sites
 Unexposed sites

Minimal or no trauma

Yes No

Ever required medical attention ?

Yes No

if yes, please specify

.....

Consultation only

Yes No

Notes

.....

.....

MOST SEVERE PRESENTATION

Type

- Petechiae
 Bruises
 Hematomas

Location of bruises (if any)

- Exposed sites
 Unexposed sites

Minimal or no trauma

Yes No

Ever required medical attention ?

Yes No

if yes, please specify

.....

Consultation only

Yes No

Notes

.....

.....

Bleeding from minor wounds

No

Trivial

N.E. Yes

If yes, please fill in the following boxes

AVERAGE PRESENTATION

Number episodes/year

- less than 1
- 1 - 5
- 6 - 12
- more than 12

Duration of average single episode (min.)

- one to ten minutes
- more than ten minutes

Ever required medical attention ?

Yes

No

if yes, please specify

Surgical hemostasis

Blood transfusion

Notes

.....
.....

MOST SEVERE PRESENTATION

Duration of episode (min.)

- one to ten minutes
- more than ten minutes

Required medical attention?

Yes

No

if yes, please specify

Surgical hemostasis

Blood transfusion

Notes

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.....

Oral cavity bleeding	No <input type="checkbox"/>	Trivial <input type="checkbox"/>	N.E. <input type="checkbox"/>	Yes <input type="checkbox"/>
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If yes, please fill in the following boxes

AVERAGE PRESENTATION	
Type of bleeding	<input type="checkbox"/> Tooth eruption <input type="checkbox"/> Gums, spontaneous <input type="checkbox"/> Gums, after brushing <input type="checkbox"/> Bites to lip & tongue
Ever required medical attention ?	Yes <input type="checkbox"/> No <input type="checkbox"/>
if yes, please specify	
Consultation only	<input type="checkbox"/>
Surgical hemostasis	<input type="checkbox"/>
Blood transfusion	<input type="checkbox"/>
Notes

MOST SEVERE PRESENTATION	
Type of bleeding	<input type="checkbox"/> Tooth eruption <input type="checkbox"/> Gums, spontaneous <input type="checkbox"/> Gums, after brushing <input type="checkbox"/> Bites to lip & tongue
Required medical attention?	Yes <input type="checkbox"/> No <input type="checkbox"/>
if yes, please specify	
Consultation only	<input type="checkbox"/>
Surgical hemostasis	<input type="checkbox"/>
Blood transfusion	<input type="checkbox"/>
Notes

Gastrointestinal bleeding	No <input type="checkbox"/>	Trivial <input type="checkbox"/>	N.E. <input type="checkbox"/>	Yes <input type="checkbox"/>
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If yes, please fill in the following boxes

AVERAGE PRESENTATION	
Number of episodes	<input type="text"/> <input type="text"/>
Type of bleeding	<input type="checkbox"/> Hematemesis <input type="checkbox"/> Melena <input type="checkbox"/> Hematochezia
Presence of associated GI disease	Yes <input type="checkbox"/> No <input type="checkbox"/> Specify: <input type="checkbox"/> Ulcer <input type="checkbox"/> Portal hypertension <input type="checkbox"/> Angiodysplasia
Ever required medical attention ?	Yes <input type="checkbox"/> No <input type="checkbox"/>
if yes, please specify	
Consultation only	<input type="checkbox"/>
Surgical hemostasis	<input type="checkbox"/>
Blood transfusion	<input type="checkbox"/>
Notes

MOST SEVERE PRESENTATION	
Type of bleeding	<input type="checkbox"/> Hematemesis <input type="checkbox"/> Melena <input type="checkbox"/> Hematochezia
Presence of associated GI disease	Yes <input type="checkbox"/> No <input type="checkbox"/> Specify: <input type="checkbox"/> Ulcer <input type="checkbox"/> Portal hypertension <input type="checkbox"/> Angiodysplasia
Required medical attention?	Yes <input type="checkbox"/> No <input type="checkbox"/>
if yes, please specify	
Consultation only	<input type="checkbox"/>
Surgical hemostasis	<input type="checkbox"/>
Blood transfusion	<input type="checkbox"/>
Notes

Tooth extraction

Total number of tooth extractions Number of extraction followed by bleeding

Please fill in a separate box for each extraction, if any: photocopy if necessary.

Bleeding after first extraction	No <input type="checkbox"/>	Trivial <input type="checkbox"/>	N.E. <input type="checkbox"/>	Yes <input type="checkbox"/>
If yes, please complete:				
Age at extraction	<input type="text"/> <input type="text"/>	Type of extraction	<input type="checkbox"/> Deciduous <input type="checkbox"/> Permanent <input type="checkbox"/> Molar	
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"/> Blood transfusion			
Notes			

Bleeding after second extraction	No <input type="checkbox"/>	Trivial <input type="checkbox"/>	N.E. <input type="checkbox"/>	Yes <input type="checkbox"/>
If yes, please complete:				
Age at extraction	<input type="text"/> <input type="text"/>	Type of extraction	<input type="checkbox"/> Deciduous <input type="checkbox"/> Permanent <input type="checkbox"/> Molar	
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"/> Blood transfusion			
Notes			

Bleeding after third extraction	No <input type="checkbox"/>	Trivial <input type="checkbox"/>	N.E. <input type="checkbox"/>	Yes <input type="checkbox"/>
If yes, please complete:				
Age at extraction	<input type="text"/> <input type="text"/>	Type of extraction	<input type="checkbox"/> Deciduous <input type="checkbox"/> Permanent <input type="checkbox"/> Molar	
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"/> Blood transfusion			
Notes			

MOST SEVERE BLEEDING AFTER 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	
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"/> Blood transfusion			
Notes			

Surgery

Total number of surgeries Number of surgeries followed by bleeding

Please fill in a separate box for each extraction, if any: photocopy if necessary.

Bleeding after first surgery	No <input type="checkbox"/>	Trivial <input type="checkbox"/>	N.E. <input type="checkbox"/>	Yes <input type="checkbox"/>
If yes, please complete:				
Age at surgery	<input type="text"/>	<input type="text"/>	Type of surgery	<input type="checkbox"/> Major-abdominal <input type="checkbox"/> Major-thoracic <input type="checkbox"/> Major-gynecology <input type="checkbox"/> Other
			<input type="checkbox"/> Tonsillectomy/Adenoids <input type="checkbox"/> Pharynx/Nose	
Bleeding after surgery?		Yes <input type="checkbox"/>		No <input type="checkbox"/>
Actions taken to control bleeding		<input type="checkbox"/> None <input type="checkbox"/> Resuturing/surgical <input type="checkbox"/> Blood transfusion <input type="checkbox"/> Other		
Notes			

Bleeding after second surgery	No <input type="checkbox"/>	Trivial <input type="checkbox"/>	N.E. <input type="checkbox"/>	Yes <input type="checkbox"/>
If yes, please complete:				
Age at surgery	<input type="text"/>	<input type="text"/>	Type of surgery	<input type="checkbox"/> Major-abdominal <input type="checkbox"/> Major-thoracic <input type="checkbox"/> Major-gynecology <input type="checkbox"/> Other
			<input type="checkbox"/> Tonsillectomy/Adenoids <input type="checkbox"/> Pharynx/Nose	
Bleeding after surgery?		Yes <input type="checkbox"/>		No <input type="checkbox"/>
Actions taken to control bleeding		<input type="checkbox"/> None <input type="checkbox"/> Resuturing/surgical <input type="checkbox"/> Blood transfusion <input type="checkbox"/> Other		
Notes			

Bleeding after third surgery	No <input type="checkbox"/>	Trivial <input type="checkbox"/>	N.E. <input type="checkbox"/>	Yes <input type="checkbox"/>
If yes, please complete:				
Age at surgery	<input type="text"/> <input type="text"/>	Type of surgery	<input type="checkbox"/> Major-abdominal <input type="checkbox"/> Major-thoracic <input type="checkbox"/> Major-gynecology <input type="checkbox"/> Other	
		<input type="checkbox"/> Tonsillectomy/Adenoids <input type="checkbox"/> Pharynx/Nose		
Bleeding after surgery?	Yes <input type="checkbox"/>	No <input type="checkbox"/>		
Actions taken to control bleeding	<input type="checkbox"/> None <input type="checkbox"/> Resuturing/surgical <input type="checkbox"/> Blood transfusion <input type="checkbox"/> Other			
Notes			

MOST SEVERE BLEEDING AFTER SURGERY				
Age at surgery	<input type="text"/> <input type="text"/>	Type of surgery	<input type="checkbox"/> Major-abdominal <input type="checkbox"/> Major-thoracic <input type="checkbox"/> Major-gynecology <input type="checkbox"/> Other	
		<input type="checkbox"/> Tonsillectomy/Adenoids <input type="checkbox"/> Pharynx/Nose		
Bleeding after surgery?	Yes <input type="checkbox"/>	No <input type="checkbox"/>		
Actions taken to control bleeding	<input type="checkbox"/> None <input type="checkbox"/> Resuturing/surgical <input type="checkbox"/> Blood transfusion <input type="checkbox"/> Other			
Notes			

Menorrhagia

Duration of average menstruation (days)

Duration of heavy days

Ever suffered from menorrhagia ?

No

Trivial

N.E.

Yes

If yes, please fill in the following boxes

AVERAGE PRESENTATION

Age of maximum severity

14 - 25

26 - 35

more than 36

Ever required medical attention ?

Yes

No

if yes, please specify

Consultation only

Pill use

Dilatation & curettage

Hysterectomy

Blood transfusion

Iron therapy

Notes

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.....

MOST SEVERE PRESENTATION

Age of bleeding

14 - 25

26 - 35

more than 36

Required medical attention?

Yes

No

if yes, please specify

Consultation only

Pill use

Dilatation & curettage

Hysterectomy

Blood transfusion

Iron therapy

Notes

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Post-partum hemorrhage

Total number of deliveries Number of deliveries followed by bleeding

Please fill in a separate box for each delivery, if any; photocopy if necessary

Bleeding after first delivery	No <input type="checkbox"/>	Trivial <input type="checkbox"/>	N.E. <input type="checkbox"/>	Yes <input type="checkbox"/>
If yes, complete:				
Ever required medical attention ?		Yes <input type="checkbox"/>		No <input type="checkbox"/>
if yes, please specify				
Dilatation & curettage		<input type="checkbox"/>		
Hysterectomy		<input type="checkbox"/>		
Blood transfusion		<input type="checkbox"/>		
Iron therapy		<input type="checkbox"/>		
Other		<input type="checkbox"/>		
Notes			

Bleeding after second delivery	No <input type="checkbox"/>	Trivial <input type="checkbox"/>	N.E. <input type="checkbox"/>	Yes <input type="checkbox"/>
If yes, complete:				
Ever required medical attention ?		Yes <input type="checkbox"/>		No <input type="checkbox"/>
if yes, please specify				
Dilatation & curettage		<input type="checkbox"/>		
Hysterectomy		<input type="checkbox"/>		
Blood transfusion		<input type="checkbox"/>		
Iron therapy		<input type="checkbox"/>		
Other		<input type="checkbox"/>		
Notes			

Bleeding after third delivery	No <input type="checkbox"/>	Trivial <input type="checkbox"/>	N.E. <input type="checkbox"/>	Yes <input type="checkbox"/>
If yes, complete:				
Ever required medical attention ?	Yes <input type="checkbox"/>	No <input type="checkbox"/>		
if yes, please specify				
Dilatation & curettage	<input type="checkbox"/>			
Hysterectomy	<input type="checkbox"/>			
Blood transfusion	<input type="checkbox"/>			
Iron therapy	<input type="checkbox"/>			
Other	<input type="checkbox"/>			
Notes			

MOST SEVERE BLEEDING AFTER DELIVERY				
Required medical attention?	Yes <input type="checkbox"/>	No <input type="checkbox"/>		
if yes, please specify				
Dilatation & curettage	<input type="checkbox"/>			
Hysterectomy	<input type="checkbox"/>			
Blood transfusion	<input type="checkbox"/>			
Iron therapy	<input type="checkbox"/>			
Other	<input type="checkbox"/>			
Notes			

Other bleedings	No <input type="checkbox"/>	Trivial <input type="checkbox"/>	N.E. <input type="checkbox"/>	Yes <input type="checkbox"/>
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If yes, please fill in the following boxes

AVERAGE PRESENTATION	
Please specify type of bleeding
Ever required medical attention ?	Yes <input type="checkbox"/> No <input type="checkbox"/>
if yes, please specify	
Blood transfusion	<input type="checkbox"/>
Desmopressin	<input type="checkbox"/>
Replacement therapy	<input type="checkbox"/>
Iron therapy	<input type="checkbox"/>
Antifibrinolytics	<input type="checkbox"/>
Notes

MOST SEVERE PRESENTATION	
Please specify type of bleeding
Required medical attention ?	Yes <input type="checkbox"/> No <input type="checkbox"/>
if yes, please specify	
Blood transfusion	<input type="checkbox"/>
Desmopressin	<input type="checkbox"/>
Replacement therapy	<input type="checkbox"/>
Iron therapy	<input type="checkbox"/>
Antifibrinolytics	<input type="checkbox"/>
Notes

Muscle hematomas or hemarthrosis	No <input type="checkbox"/>	Trivial <input type="checkbox"/>	N.E. <input type="checkbox"/>	Yes <input type="checkbox"/>
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If yes, please fill in the following boxes

AVERAGE PRESENTATION	
Please specify type & location
Spontaneous ?	Yes <input type="checkbox"/> No <input type="checkbox"/>
Ever required medical attention ?	Yes <input type="checkbox"/> No <input type="checkbox"/>
If yes, please specify	
Blood transfusion	<input type="checkbox"/>
Desmopressin	<input type="checkbox"/>
Replacement therapy	<input type="checkbox"/>
Notes

MOST SEVERE PRESENTATION	
Please specify type & location
Spontaneous ?	Yes <input type="checkbox"/> No <input type="checkbox"/>
Required medical attention?	Yes <input type="checkbox"/> No <input type="checkbox"/>
If yes, please specify	
Blood transfusion	<input type="checkbox"/>
Desmopressin	<input type="checkbox"/>
Replacement therapy	<input type="checkbox"/>
Notes

APPENDIX

Calculation of the individual BLEEDING SCORE

A symptom-specific grade, ranging from 0 to 3 (see following Table), is attributed to each bleeding symptom reported in the Questionnaire. The sum of all grades within an individual is the Bleeding Score. For each symptom, the most severe occurrence contributed to the Bleeding Score.

Symptoms	Assigned score	
Epistaxis	0 = no or trivial 1 = present	2 = packing, cauterization, 3 = transfusion, replacement
Cutaneous symptoms	0 = no or trivial 1 = petechiae or bruises	2 = hematomas 3 = medical consultation
Minor wounds	0 = no or trivial 1 = present (1-5 episodes/year)	2 = medical attention 3 = surgery / blood transfusion
Oral cavity bleeding	0 = no or trivial 1 = present	2 = medical attention 3 = surgery / blood transfusion
Gastrointestinal bleeding	0 = no or trivial 1 = present	2 = medical attention 3 = surgery / blood transfusion
Post-partum hemorrhage	0 = no or trivial 1 = present, iron therapy	2 = blood transfusion, dilatation-curettage, suturing 3 = hysterectomy
Muscle hematomas or hemarthrosis	0 = no or trivial 1 = present	2 = medical attention 3 = transfusion, intervention
Tooth extraction (most severe episode)	0 = no or trivial 1 = present	2 = suturing or packing 3 = transfusion
Surgery (most severe episode)	0 = no or trivial 1 = present	2 = suturing or resurgery 3 = transfusion
Menorrhagia	0 = no or trivial 1 = present	2 = consultation, pill use, iron therapy 3 = transfusion, hysterectomy, dilatation-curettage, replacement therapy