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

CONTENT:

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, Tosetto A, Batlle 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 interntional, 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:

Francesco Rodeghiero – email: rodeghiero@hemato.ven.it

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 □	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 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 □
Seasonal correlation	Yes □	No □
Cessation	□spontaneous □after short compression □by medical intervention	
Age of maximum severity	□ less than 14 years □ 14 to 45 years □ more than 45 years	
Ever required medical attention ?	Yes □	No □
if yes, please specify		
Consultation only	□	
Cauterization	□	
Packing	□	
Blood transfusion	□	
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	…… …… ……	

Cutaneous symptoms	No □	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	…… ……	

Oral cavity bleeding	No □	Trivial □	N.E. □	Yes □

If yes, please fill in the following boxes

AVERAGE PRESENTATION

Type of bleeding	□ Tooth eruption □ Gums, spontaneous □ Gums, after brushing □ Bites to lip & tongue	
Ever required medical attention ?	Yes □	No □
if yes, please specify		
Consultation only	□	
Surgical hemostasis	□	
Blood transfusion	□	
Notes	…… ……	

MOST SEVERE PRESENTATION

Type of bleeding	□ Tooth eruption □ Gums, spontaneous □ Gums, after brushing □ Bites to lip & tongue	
Required medical attention?	Yes □	No □
if yes, please specify		
Consultation only	□	
Surgical hemostasis	□	
Blood transfusion	□	
Notes	…… ……	

Gastrointestinal bleeding	No □	Trivial □	N.E. □	Yes □

If yes, please fill in the following boxes

AVERAGE PRESENTATION

Number of episodes	□□	
Type of bleeding	□ Hematemesis □ Melena □ Hematochezia	
Presence of associated GI disease	Yes □ Specify: □ Ulcer □ Portal hypertension □ Angiodysplasia	No □
Ever required medical attention ?	Yes □	No □
if yes, please specify Consultation only Surgical hemostasis Blood transfusion	 □ □ □	
Notes	…… ……	

MOST SEVERE PRESENTATION

Type of bleeding	□ Hematemesis □ Melena □ Hematochezia	
Presence of associated GI disease	Yes □ Specify: □ Ulcer □ Portal hypertension □ Angiodysplasia	No □
Required medical attention?	Yes □	No □
if yes, please specify Consultation only Surgical hemostasis Blood transfusion	 □ □ □	
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 ☐ Trivial ☐ N.E. ☐ Yes ☐

If yes, please complete:

Age at extraction ☐☐ Type of extraction ☐ Deciduous ☐ Permanent ☐ Molar

Bleeding after extraction? Yes ☐ No ☐

Actions taken to control bleeding ☐ None ☐ Resuturing ☐ Packing ☐ Blood transfusion

Notes ……
……

Bleeding after second extraction No ☐ Trivial ☐ N.E. ☐ Yes ☐

If yes, please complete:

Age at extraction ☐☐ Type of extraction ☐ Deciduous ☐ Permanent ☐ Molar

Bleeding after extraction? Yes ☐ No ☐

Actions taken to control bleeding ☐ None ☐ Resuturing ☐ Packing ☐ Blood transfusion

Notes ……
……

Bleeding after third extraction	No □	Trivial □	N.E. □	Yes □
If yes, please complete:				
Age at extraction	□□	Type of extraction	□ Deciduous □ Permanent □ Molar	
Bleeding after extraction?		Yes □	No □	
Actions taken to control bleeding		□ None □ Resuturing □ Packing □ Blood transfusion		
Notes		……		

MOST SEVERE BLEEDING AFTER TOOTH EXTRACTION			
Age at extraction	□□	Type of extraction	□ Deciduous □ Permanent □ Molar
Bleeding after extraction?		Yes □	No □
Actions taken to control bleeding		□ None □ Resuturing □ Packing □ 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 ☐ Trivial ☐ N.E. ☐ Yes ☐

If yes, please complete:

Age at surgery ☐☐

Type of surgery

☐ Tonsillectomy/Adenoids
☐ Pharynx/Nose
☐ Major-abdominal
☐ Major-thoracic
☐ Major-gynecology
☐ Other

Bleeding after surgery? Yes ☐ No ☐

Actions taken to control bleeding

☐ None
☐ Resuturing/surgical
☐ Blood transfusion
☐ Other

Notes

……..
……..

Bleeding after second surgery No ☐ Trivial ☐ N.E. ☐ Yes ☐

If yes, please complete:

Age at surgery ☐☐

Type of surgery

☐ Tonsillectomy/Adenoids
☐ Pharynx/Nose
☐ Major-abdominal
☐ Major-thoracic
☐ Major-gynecology
☐ Other

Bleeding after surgery? Yes ☐ No ☐

Actions taken to control bleeding

☐ None
☐ Resuturing/surgical
☐ Blood transfusion
☐ Other

Notes

……..
……..

Bleeding after third surgery	No □	Trivial □	N.E. □	Yes □

If yes, please complete:

Age at surgery □□

Type of surgery

□ Tonsillectomy/Adenoids
□ Pharynx/Nose
□ Major-abdominal
□ Major-thoracic
□ Major-gynecology
□ Other

Bleeding after surgery? Yes □ No □

Actions taken to control bleeding

□ None
□ Resuturing/surgical
□ Blood transfusion
□ Other

Notes

……..

……..

MOST SEVERE BLEEDING AFTER SURGERY

Age at surgery □□

Type of surgery

□ Tonsillectomy/Adenoids
□ Pharynx/Nose
□ Major-abdominal
□ Major-thoracic
□ Major-gynecology
□ Other

Bleeding after surgery? Yes □ No □

Actions taken to control bleeding

□ None
□ Resuturing/surgical
□ Blood transfusion
□ Other

Notes

……..

……..

Menhorragia

Duration of average menstruation (days) ☐☐ Duration of heavy days ☐☐

Ever suffered from menhorragia ? 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 ……
……

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 ……
……

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 ☐ Trivial ☐ N.E. ☐ Yes ☐

If yes, complete:

Ever required medical attention ? Yes ☐ No ☐

if yes, please specify

Dilatation & curettage ☐
Hysterectomy ☐
Blood transfusion ☐
Iron therapy ☐
Other ☐

Notes ……
……

Bleeding after second delivery No ☐ Trivial ☐ N.E. ☐ Yes ☐

If yes, complete:

Ever required medical attention ? Yes ☐ No ☐

if yes, please specify

Dilatation & curettage ☐
Hysterectomy ☐
Blood transfusion ☐
Iron therapy ☐
Other ☐

Notes ……
……

Bleeding after third delivery No ☐ Trivial ☐ N.E. ☐ Yes ☐

If yes, complete:

Ever required medical attention ? Yes ☐ No ☐

if yes, please specify

Dilatation & curettage ☐
Hysterectomy ☐
Blood transfusion ☐
Iron therapy ☐
Other ☐

Notes ……
……

MOST SEVERE BLEEDING AFTER DELIVERY

Required medical attention? Yes ☐ No ☐

if yes, please specify

Dilatation & curettage ☐
Hysterectomy ☐
Blood transfusion ☐
Iron therapy ☐
Other ☐

Notes ……
……

Other bleedings	No □	Trivial □	N.E. □	Yes □

If yes, please fill in the following boxes

AVERAGE PRESENTATION

Please specify type of bleeding . . . . . .

. . . . . .

Ever required medical attention ? Yes □ No □

if yes, please specify

Blood transfusion □

Desmopressin □

Replacement therapy □

Iron therapy □

Antifibrinolytics □

Notes ……
……

MOST SEVERE PRESENTATION

Please specify type of bleeding . . . . . .

. . . . . .

Required medical attention ? Yes □ No □

if yes, please specify

Blood transfusion □

Desmopressin □

Replacement therapy □

Iron therapy □

Antifibrinolytics □

Notes ……
……

Muscle hematomas or hemarthrosis	No □	Trivial □	N.E. □	Yes □

If yes, please fill in the following boxes

AVERAGE PRESENTATION

Please specify type & location . . . . . .

. . . . . .

Spontaneous ? Yes □ No □

Ever required medical attention ? Yes □ No □

If yes, please specify

Blood transfusion □

Desmopressin □

Replacement therapy □

Notes ……
……..

MOST SEVERE PRESENTATION

Please specify type & location . . . . . .

. . . . . .

Spontaneous ? Yes □ No □

Required medical attention? Yes □ No □

If yes, please specify

Blood transfusion □

Desmopressin □

Replacement therapy □

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