

## ORIGINAL ARTICLE

# Generation and validation of the Condensed MCMDM-1VWD Bleeding Questionnaire for von Willebrand disease

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**Summary.** *Background:* Given the challenges involved in obtaining accurate bleeding histories, attempts at standardization have occurred and the value of quantifying hemorrhagic symptoms has been recognized. *Patients/methods:* An extensive validated bleeding questionnaire (MCMDM-1VWD) was condensed by eliminating all details that did not directly affect the bleeding score (BS) and the correlation between the two versions was tested. Additionally, the diagnostic utility of the condensed version was prospectively tested. *Results:* Data on 259 individuals who were administered the questionnaire are presented here; 217 being prospectively investigated for von Willebrand disease (VWD) (group 1) and 42 previously known to have type 1, 2 or 3 VWD (group 2). Of the 217 prospectively investigated, 35 had positive BS ( $\geq 4$ ) and 182 had negative scores. Seven individuals (all with positive BS) had laboratory results consistent with type 1 VWD. This results in a sensitivity of 100% and a specificity of 87%. The positive predictive value is 0.20 and the negative predictive value is 1. The correlation between the full MCMDM-1VWD and condensed versions is excellent (Spearman's 0.97,  $P < 0.001$ , linear regression  $r^2 = 96.4$ ). Inter-observer reliability for the condensed version is reasonable (Spearman's 0.72,  $P < 0.001$  and intra-class correlation coefficient 0.805,  $P < 0.001$ ). There was a significant difference in BS between subtypes of VWD, with type 3  $\gg$  type 2  $\gg$  type 1 VWD (ANOVA  $P < 0.001$ ). There is a strong inverse relationship between VWF:Ag level and BS (Spearman's  $-0.411$ ,  $P < 0.001$ ). *Conclusions:* The Condensed MCMDM-1VWD Bleeding Questionnaire is an efficient, effective tool in the evaluation of patients for VWD.

**Keywords:** bleeding questionnaire, bleeding score, VWD.

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## Introduction

The accurate assessment of hemorrhagic symptoms is a well-recognized challenge for both patients and physicians, but forms a critical component in the diagnosis of a number of mild bleeding disorders, including von Willebrand disease (VWD). Bleeding histories are subjective and significant symptoms can be overlooked and interpreted as normal. In contrast, minimal or trivial symptoms may be given undue consideration, especially given the high frequency of bleeding symptoms reported by the general population [1]. Many attempts at standardizing bleeding histories have been made in an effort to both prevent unwarranted laboratory testing and to prevent false-positive diagnoses and the value of a quantitative scoring system has been recognized.

Quantitation of bleeding symptoms has many potential advantages over a binary classification system including facilitating the exploration of variability in bleeding severity, informing treatment decisions and providing an efficient means of communication between health care providers. The quantitative bleeding questionnaire that is detailed on the ISTH website ([http://www.med.unc.edu/isth/ssc/collaboration/Bleeding\\_Type1\\_VWD.pdf](http://www.med.unc.edu/isth/ssc/collaboration/Bleeding_Type1_VWD.pdf)) was developed and validated by a group of investigators in Vicenza, Italy [2]. This detailed questionnaire is 17 pages long and takes approximately 40 min to administer. After administration of this questionnaire, the bleeding score is generated by summing the severity of all bleeding symptoms reported by a subject. Bleeding symptoms are scored from 0 (absence or trivial symptoms) to 3 (symptom requiring medical intervention).

In order to improve the sensitivity and specificity of the bleeding score, this scoring system was later revised to increase the range of possible grades from  $-1$  (absence of bleeding after significant hemostatic challenge such as two dental extractions or surgeries) to 4 (symptoms requiring the most significant medical intervention such as infusion of clotting factor concentrates or surgery to control bleeding). This  $-1$  to 4 version was used for the European MCMDM-1VWD Study and discriminated effectively between type 1 VWD and unaffected individuals [3]. We condensed this version of the

questionnaire by maintaining only the details that directly affect the bleeding score resulting in a six-page questionnaire that can be administered in 5–10 min. We compared the bleeding score with laboratory criteria defining type 1 VWD to determine the diagnostic utility of the Condensed MCMDM-1VWD Bleeding Questionnaire, and also administered the questionnaire to a group of individuals previously known to have VWD.

## Patients, materials and methods

### Patients

To determine the normal range of bleeding scores, healthy subjects with no known problem with bleeding or bruising were recruited from community volunteers who responded to advertisements posted on the campus of Queen's University in Kingston, Ontario.

Additional subjects were consecutively recruited from the waiting rooms of either primary or tertiary medical clinics and were from one of two groups: subjects in group 1 were being prospectively investigated for VWD (unrelated subjects recruited from primary care clinics) and group 2 comprised of subjects previously known to have type 1, 2 or 3 VWD (recruited from a tertiary care medical clinic). The Condensed MCMDM-1VWD Bleeding Questionnaire was administered and blood work was taken as detailed below. A subset also had the full MCMDM-1VWD questionnaire administered and a different subset had the condensed version administered twice, by two different observers. All subjects gave informed consent and Research Ethics Board approval was obtained from Queen's University.

### Generation of the Condensed MCMDM-1VWD Bleeding Questionnaire

In order to condense the MCMDM-1VWD Bleeding Questionnaire only the details that directly affect the bleeding score were maintained. Additionally, a few small changes in wording were made as shown in the scoring table at the end of the bleeding questionnaire (included as a supplemental file).

### Laboratory testing

Venous blood samples were collected by phlebotomy (on the same day that the questionnaire was administered) in 3.2% sodium citrate [at a ratio of 9:1 (v/v)] and EDTA. ABO blood group, von Willebrand factor (VWF):Ag, VWF:RCo and FVIII:C were performed on all subjects with positive bleeding scores. The VWF tests were repeated on at least one other occasion for this group and mean data are presented. All tests were also performed on those with negative bleeding scores with the exception of VWF:RCo which was performed only if the VWF:Ag < 0.80 U mL<sup>-1</sup>. Additionally, these subjects only had one set of VWF tests performed. VWF multimers were performed if either the VWF:Ag or VWF:RCo were less

than 0.50 U mL<sup>-1</sup>. ABO blood group and VWF tests were performed according to previously published methods [4].

### Laboratory definition of von Willebrand disease

The laboratory definitions that were used are those that are commonly used within our group both in the clinical and research settings [4–6]. For type 1 VWD, this includes a VWF:Ag and VWF:RCo between 0.05 and 0.50 IU mL<sup>-1</sup> on at least two occasions, a RCo:Ag ratio > 0.60 and normal VWF multimers. For type 2A, abnormal lab investigations include a VWF:Ag and/or VWF:RCo between 0.05 and 0.50 IU mL<sup>-1</sup> on at least two occasions and abnormal multimers and type 2M is defined as VWF:Ag and/or VWF:RCo between 0.05 and 0.50 IU mL<sup>-1</sup> on at least two occasions, RCo:Ag ratio < 0.60 and normal multimers. Type 2B VWD is defined the same as type 2A VWD with the additional requirement of a ristocetin induced platelet aggregation (RIPA) with increased sensitivity. Type 3 VWD is defined as VWF:Ag and/or VWF:RCo < 0.05 IU mL<sup>-1</sup> and FVIII:C < 0.10 IU mL<sup>-1</sup>.

## Results

### Patients and bleeding scores

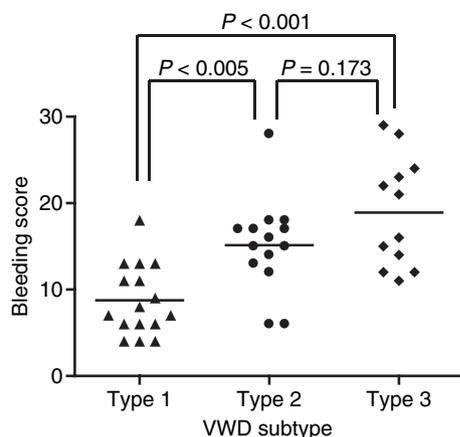
A total of 100 healthy subjects (no known problem with bleeding or bruising) were recruited in order to determine the range of normal bleeding scores. There were 35 males and 65 females included, with a mean age of 39 years (range 20–88 years). The mean bleeding score was 0.16 with a standard deviation (SD) of 1.7, therefore the range of normal bleeding scores was determined to be –3.2 to +3.6 (mean ± 2 SDs given that the data were normally distributed). There was no difference in bleeding scores between genders. Bleeding scores are expressed in whole numbers, therefore a positive (or abnormal) bleeding score was considered to be ≥4 for all analyses.

Of the 259 individuals reported here, 217 were from group 1 (prospective assessment for VWD) and 42 were from group 2 (known diagnosis of VWD). With regards to group 1, 35 had positive bleeding scores leaving 182 with negative bleeding scores. Of the 35 with positive BS, seven had laboratory investigations consistent with type 1 VWD, leaving 28 with positive BS but no evidence of VWD. Fifteen out of the 28 underwent additional coagulation investigations and five had platelet function defects identified, as previously reported [7]. None of the 182 with negative bleeding scores met the criteria for type 1 VWD (Table 1). With regards to group 2, 42 subjects were included (type 1 VWD  $n = 16$ , type 2 VWD  $n = 14$ , type 3 VWD  $n = 12$ ). The type 2 VWD group comprised of nine with type 2B, four with type 2A and one with type 2M VWD. Group 2 comprised of 16 males, 26 females and the mean age is 30 years (range 4–69 years). The median BS for all of group 2 is 14 (range 4–29) and there were significant differences in BS between the subtypes of VWD with type 3 >> type 2 >> type 1 VWD (median BS for type 3 = 19,

**Table 1** Group 1 ( $n = 217$ ) which includes those that met the criteria for type 1 VWD ( $n = 7$ ), those with positive bleeding scores but normal VWF lab tests (non-VWD bleeders  $n = 28$ ) and those with negative bleeding scores ( $n = 182$ )

	Type 1 VWD ( $n = 7$ )	Non-VWD bleeders ( $n = 28$ )	Negative BS ( $n = 182$ )	<i>P</i> -value
Females (%)	6 (86)	27 (96)	123 (68)	0.027
Mean age, years (range)	35 (20–58)	42 (11–68)	45 (19–81)	0.213
Age groups, years				
< 15	0	1	0	
15–30	3	7	32	
31–50	3	11	87	
51–70	1	9	58	
> 70	0	0	5	
Blood group O (%)	5 (71)	17 (61)	85 (48)	0.333
Mean VWF:Ag, U mL <sup>-1</sup> (range)	0.48 (0.44–0.49)	1.28 (0.64–2.46)	1.17 (0.52–2.96)	0.001
Mean VWF:RCO, U mL <sup>-1</sup> (range)	0.37 (0.28–0.44)	0.97 (0.52–1.91)	0.80 (0.27–1.76)	< 0.001
Mean FVIII:C, U mL <sup>-1</sup> (range)	0.83 (0.47–1.27)	1.24 (0.67–1.83)	1.26 (0.48–2.75)	0.026
Median BS (range)	10 (5–15)	6 (4–21)	-1 (-3 to 3)	< 0.001

BS, bleeding score; VWD, von Willebrand disease; VWF, von Willebrand factor. In the negative BS column, the range of VWF:RCO includes values < 0.50 IU mL<sup>-1</sup> from three individuals. All had normal VWF:Ag levels (and therefore did not meet the criteria for type 1 VWD) and negative bleeding scores and all are blood group O. Subject characteristics were compared using chi-square tests for categorical data and one-way analysis of variance (ANOVA) for linear data. The non-parametric Kruskal–Wallis test was used to compare the bleeding scores as they were not normally distributed. Tukey's *post-hoc* testing, pair-wise chi-square testing and the Mann–Whitney *U*-test were used to further examine between-group differences. Gender: VWD vs. non-VWD bleeders  $P = 0.876$ , VWD vs. negative BS  $P = 0.585$ , non-VWD bleeders vs. negative BS  $P = 0.028$ . VWF:Ag: VWD vs. Non-VWD bleeders  $P = 0.002$ , VWD vs. negative BS  $P = 0.001$ , non-VWD bleeders vs. negative BS  $P = 0.813$ . VWF:RCO: VWD vs. non-VWD bleeders  $P < 0.001$ , VWD vs. negative BS  $P < 0.001$ , non-VWD bleeders vs. negative BS  $P = 0.028$ . FVIII: VWD vs. non-VWD bleeders  $P = 0.039$ , VWD vs. negative BS  $P = 0.020$ , non-VWD bleeders vs. negative BS  $P = 0.991$ . BS: VWD vs. non-VWD bleeders  $P = 0.755$ , VWD vs. negative BS  $P < 0.001$ , non-VWD bleeders vs. negative BS  $P < 0.001$ .



**Fig. 1.** Shows the 42 subjects included in group 2. Median bleeding score is shown in the horizontal dark bar by diagnosis – type 1, 2 and 3 von Willebrand disease (VWD). The overall comparison was significant (ANOVA  $P < 0.001$  – not shown on figure). Between-group analysis was performed using a *post-hoc* Tukey's (*P*-values shown on figure).

Type 2 = 16, type 1 = 8) (ANOVA  $P < 0.001$ , *post hoc* Tukey's T1/T2  $P = 0.005$ , T1/T3  $P < 0.001$ , T2/T3  $P = 0.173$ ) (Fig. 1).

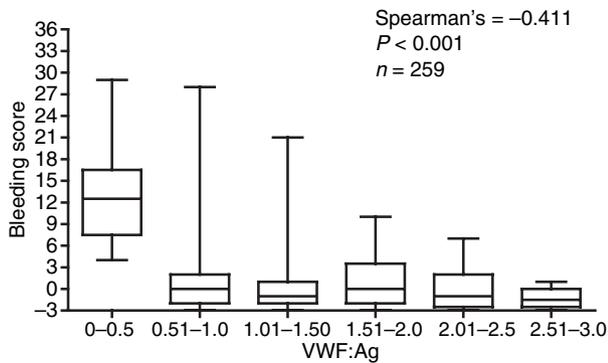
#### Analysis of the Condensed MCMDM-1VWD Bleeding Questionnaire

Analysis of the 217 subjects included in group 1 shows that the sensitivity, specificity, positive predictive value (PPV) and negative predictive value (NPV) of the Condensed MCMDM-1VWD Bleeding Questionnaire for type 1 VWD are 100%,

87%, 0.20 and 1, respectively. Seventeen individuals with negative bleeding scores have subsequently undergone major surgery (surgery involving laparotomy, thoracotomy or total joint replacement) and none experienced any significant bleeding. The likelihood ratio for a positive BS ( $\geq 4$ ) is 7.5 (95% confidence limits 5.3–10.5). The receiver operator characteristic (ROC) analysis shows an area under the curve of 0.96 ( $P < 0.001$ ) which means that the questionnaire can very accurately distinguish between affected and unaffected individuals. The correlation between the full MCMDM-1VWD Bleeding Questionnaire and our condensed version (calculated on a subset of 17 subjects administered both versions approximately 2 weeks apart) is excellent producing a Spearman's rho of 0.97 ( $P < 0.001$ ) and an  $r^2 = 96.4$  from the linear regression. Inter-observer reliability (calculated on a subset of 24 subjects administered the condensed version by two different observers approximately 3 months apart) was reasonable, producing a Spearman's rho of 0.72 ( $P < 0.001$ ) and an intra-class correlation coefficient of 0.805 ( $P < 0.001$ ). When all subjects from group 1 and group 2 were included there was a strong inverse correlation between the bleeding score and the VWF:Ag level (Spearman's correlation  $-0.411$ ,  $P < 0.001$ ) (Fig. 2).

#### Discussion

Accurate assessment of hemorrhagic symptoms is a key component in the diagnosis of many mild bleeding disorders including VWD, and many attempts to standardize bleeding histories have been made [8–13]. This Condensed MCMDM-1VWD Bleeding Questionnaire provides the ability to efficiently



**Fig. 2.** Shows the strong inverse relationship between bleeding score (BS) and von Willebrand factor (VWF) level (VWF:Ag expressed as IU mL<sup>-1</sup>). All subjects are included in this figure (groups 1 and 2). The middle bar represents the median with the rectangle representing the 25–75 percentiles. The error bars represent the range.

calculate the bleeding score of an individual patient and is based on a validated and well-recognized scoring system. Two main issues with regards to our data merit comment: the lack of pediatric data and the clinical applicability of this questionnaire to a new patient presenting with hemorrhagic symptoms.

With regards to the issue of a lack of pediatric data, a review of Table 1 highlights the fact that the population reported here is largely an adult one. Furthermore, the issues specific to pediatric bleeding (including the different patterns of bleeding experienced and the lack of hemostatic challenges in younger children) require a carefully considered approach. With this in mind, a bleeding questionnaire based on the MCMDM-1VWD scoring system which includes pediatric-specific bleeding symptoms has been generated and forms the basis of an ongoing study.

There is a desire amongst clinicians who evaluate patients with hemorrhagic symptoms for a bleeding questionnaire that would be widely applicable; however, the prospective validation of such a tool for all mild bleeding disorders would be very difficult, especially for platelet function disorders. There is an acknowledged lack of standardization in the laboratory approach to platelet aggregation testing, which would affect the general applicability of any conclusions [14].

We acknowledge that in some settings, administering the full MCMDM-1VWD Bleeding Questionnaire may be more appropriate. Certainly, the additional detail gathered using that tool may be valuable, particularly in a research setting. However, we present the Condensed MCMDM-1VWD Bleeding Questionnaire as an efficient alternative; one with high diagnostic accuracy for type 1 VWD and proven reproducibility between observers. The clinical utility of our bleeding score lies in its ability to succinctly summarize a great deal of clinical information about an individual patient. This greatly aids in communication between clinicians and also could help to prioritize laboratory testing. In the primary care setting, the clinical utility lies in the high NPV and perhaps its greatest value is in the identification of patients for whom VWF testing is not necessary.

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## Disclosure of Conflict of Interests

The authors state that they have no conflict of interest.

## Supporting Information

Additional Supporting Information may be found in the online version of this article:

**Table S1.** Condensed MCMDM-1VWD Bleeding Questionnaire.

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**CONDENSED MCMDM-1 BLEEDING QUESTIONNAIRE:**



**Patient Information**

Name \_\_\_\_\_

Address \_\_\_\_\_

\_\_\_\_\_

Phone Number \_\_\_\_\_ Email \_\_\_\_\_

Gender Male  Female

Age \_\_\_\_\_ Date of Birth \_\_\_\_\_ (DD/MO/YYYY)

Ethnic Background \_\_\_\_\_

Presenting complaint of bleeding or bruising today Yes  No

Personal history of bleeding or bruising Yes  No

Ever been diagnosed with a bleeding disorder? Yes  No

Diagnosis: \_\_\_\_\_

Family history of bleeding (at least one family member) Yes  No

If yes, what was the diagnosis? \_\_\_\_\_

Pedigree:

Are you currently taking Oral Contraceptive Pills? Yes  No

If yes, brand name \_\_\_\_\_

Are you pregnant? \_\_\_\_\_ Gestation time \_\_\_\_\_

Specify any herbals and/or medications that you have taken in the past 30 days:

Name	Dose	Route	Frequency	Duration
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\_\_\_\_\_

\_\_\_\_\_

\_\_\_\_\_

\_\_\_\_\_

**Nosebleeds**

Yes

No

Number of episodes/year

< 1

6 - 12

1 - 5

> 12

Duration of average episode

< 1 minute

1 - 10 minutes

> 10 minutes

Medical attention

Yes

No

Consultation only

Cauterization/packing

Antifibrinolytics

DDAVP

Transfusion/Replacement

**Bruising**

Yes

No

Location

Exposed sites

Unexposed sites

Size of average

< 1 cm

1 - 5 cm

> 5 cm

Minimal or no trauma

Yes

No

Medical attention

Yes

No

If yes, please specify \_\_\_\_\_

**Bleeding from minor wounds**

Yes

No

- Number per year
- < 1
  - 1 – 5
  - 6 or more

- Duration of average episode
- < 5 minutes
  - > 5 minutes

- Medical attention
- Yes
  - No
- Consultation only
  - Surgical hemostasis
  - Blood transfusion/DDAVP/Replacement

**Oral cavity bleeding**

Yes

No

- Tooth eruption
- Gums, spontaneous
- Gums, after brushing
- Bites to lip and tongue

- Medical attention
- Yes
  - No
- Consultation only
  - Surgical hemostasis/Antifibrinolytic
  - Blood transfusion/DDAVP/Replacement

**Post-dental extraction**

Yes

No

- No bleeding in at least 2 extractions
- None done, or no bleeding in 1 extraction

**Medical attention**

Yes

No

- Consultation only
- Resuturing or packing
- Blood transfusion/DDAVP/Replacement

**Gastrointestinal Bleeding**

Yes

No

- Ulcer, portal hypertension, hemorrhoids
- Spontaneous
- Surgery/Blood transfusion/DDAVP/Antifibrinolytic

**Surgery**

Yes

No

- No bleeding in at least 2 surgeries
- None done, or no bleeding in 1 surgery

**Post-op medical attention**

Yes

No

- Consultation only
- Surgical hemostasis/Antifibrinolytic
- Blood transfusion/DDAVP/Replacement

**Menorrhagia**

Yes

No

Duration of average menstruation \_\_\_\_\_ days

Duration of heavy menstruation \_\_\_\_\_ days

How often do you change your pads/tampons

on heaviest days \_\_\_\_\_ hours

on average days \_\_\_\_\_ hours

What type of feminine product do you use? (i.e. panty liner, super absorbency tampon etc.)

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Medical attention  Yes

No

Consultation only

Pill use/Antifibrinolytics

Dilatation & curettage

Iron therapy

Blood transfusion/DDAVP/Replacement

Hysterectomy

**Post-partum hemorrhage**

Yes

No

No bleeding in at least 2 deliveries

No deliveries, or no bleeding in 1 delivery

Medical attention  Yes

No

Consultation only

D&C/Iron therapy/Antifibrinolytics

Blood transfusion/DDAVP/Replacement

Hysterectomy

**Muscle hematomas**

Yes

No

- Post-trauma, no therapy
- Spontaneous, no therapy
- Spontaneous or traumatic requiring DDAVP or Replacement
- Spontaneous or traumatic requiring surgical Intervention or transfusion

**Hemarthrosis**

Yes

No

- Post-trauma, no therapy
- Spontaneous, no therapy
- Spontaneous or traumatic requiring DDAVP or Replacement
- Spontaneous or traumatic requiring surgical Intervention or transfusion

**Central Nervous System Bleeding**

Yes

No

- Subdural, any intervention
- Intracerebral, any intervention

**Other**

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**Medical attention**

Yes

No

- Consultation only
- Surgical hemostasis/Antifibrinolytic
- Blood transfusion/DDAVP/Replacement

## Scoring Key

Symptom	Score	-1	0	1	2	3	4
Epistaxis	--		No or trivial (less than 5)	> 5 or more than 10'	Consultation only	Packing or cauterization or antifibrinolytic	Blood transfusion or replacement therapy or desmopressin
Cutaneous	--		No or trivial (< 1cm)	> 1 cm and no trauma	Consultation only	--	--
Bleeding from minor wounds	--		No or trivial (less than 5)	> 5 or more than 5'	Consultation only	Surgical hemostasis	Blood transfusion or replacement therapy or desmopressin
Oral cavity	--		No	Referred at least one	Consultation only	Surgical hemostasis or antifibrinolytic	Blood transfusion or replacement therapy or desmopressin
Gastrointestinal bleeding	--		No	Associated with ulcer, portal hypertension, hemorrhoids, angiodysplasia	Spontaneous	Surgical hemostasis, blood transfusion, replacement therapy, desmopressin, antifibrinolytic	--
Tooth extraction	No bleeding in at least 2 extractions	None done or no bleeding in 1 extraction	Reported, no consultation	Consultation only	Resuturing or packing	Blood transfusion or replacement therapy or desmopressin	Blood transfusion or replacement therapy or desmopressin
Surgery	No bleeding in at least 2 surgeries	None done or no bleeding in 1 surgery	Reported, no consultation	Consultation only	Surgical hemostasis or antifibrinolytic	Blood transfusion or replacement therapy or desmopressin	Blood transfusion or replacement therapy or desmopressin
Menorrhagia	--	No	Consultation only	Antifibrinolytics, pill use	Dilation & curettage, iron therapy, ablation	Blood transfusion or replacement therapy or desmopressin or hysterectomy	Hysterectomy
Postpartum hemorrhage	No bleeding in at least 2 deliveries	None done or no bleeding in 1 surgery	Consultation only	Dilation & curettage, iron therapy, antifibrinolytics	Blood transfusion or replacement therapy or desmopressin	Spontaneous or traumatic, requiring surgical intervention or blood transfusion	Spontaneous or traumatic, requiring surgical intervention or blood transfusion
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	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	Spontaneous or traumatic, requiring surgical intervention or blood transfusion
Central nervous system bleeding	--	Never	--	--	Subdural, any intervention	Intracerebral, any intervention	Intracerebral, any intervention

Scoring Key: A few small changes in wording from the original MCMDM-1 scoring key are indicated in the boxes. For tooth extraction and surgery a score of 1 was previously for “Referred in <25% of cases” and a score of 2 was previously for “Referred in >25% of cases”. For the menorrhagia category we added ablation therapy (which was not included in the original) at the same level as D&C and iron therapy.