

## **Symptomatic VWD in a Pediatric Population- a guide to administering the Bleeding Questionnaire**

### **General Points**

1. This questionnaire has been designed to be administered by a health professional asking questions of a parent/guardian and/or the child- it is not to be self-administered.
2. When administering the questionnaire the wording for each section should be "Have you ever had a problem with nosebleeds/bruising/heavy periods, etc.?"
3. A symptom must reach a particular threshold in order to be termed significant. In the following pages, under each symptom heading, a definition for "significant" is provided. If the symptom is reported by the parent/child but does not reach this threshold then it should be marked "trivial" in the appropriate box.
4. When asking questions, take care to ask "open" questions, eg. "How long does an average nosebleed last?" rather than "Do your nosebleeds last for more than ten minutes?"
5. Watch for "or" and "and" when scoring epistaxis, cutaneous and minor wound bleeding.
6. A score for "Consultation" is recorded if the patient has EVER spoken to a medical professional (doctor, nurse or dentist) about the symptom.
7. A score for "Replacement therapy" is recorded if the patient was given a product containing the haemostatic factor which is deficient. This would include platelet transfusion for platelet function disorders. "Blood transfusion" refers specifically to the transfusion of packed red cells.
8. Score the worst case for each symptom, ie. if most nosebleeds last only 5 minutes but one required cautery, the score for that category would be 3.
9. The maximum score for any symptom is 4, apart from GI which is 3 and cutaneous which is 2. For all categories, apart from the "Other" category (see below), the score is not cumulative if there have been numerous events, ie. three nosebleeds that all required cautery only score 3 for that category, not 9.
10. For the "Other" category, the most severe of each of the "other" symptoms is scored and summated to give a score for this category which may be greater than 4.

## Specific Points

### Page 1

Ethnic Background of biological mother and father should be recorded, selecting from the following list:

An aboriginal person (eg. North American Indian, Metis, Inuit, Eskimo)

White

Chinese

South Asian (eg. East Indian, Pakistani, Sri Lankan, etc.)

Black

Filipino

Latin American

Southeast Asian (eg. Cambodian, Indonesian, Laotian, Vietnamese, etc.)

Arab

West Asian (eg. Afghan, Iranian, etc.)

Japanese

Korean

Other (please specify)

For the purposes of the positive control group, the answer to the question “Presenting complaint of bleeding or bruising today?” is Yes.

First degree relative refers to parent, child or sibling of the subject and does not include half-siblings.

Remember to include the subject in the total number of 1<sup>st</sup> degree family members. In the notes section, information such as “child adopted” or “family history not known” can be added.

## Page 2

### Epistaxis

Epistaxis = Nosebleed.

Definition of significant epistaxis- Any nosebleed lasting for longer than 10 minutes OR requiring medical attention OR occurring frequently (greater than 5 times per year).

When recording the number of episodes per year, take the period of time that the symptoms were at their maximum severity.

Spontaneous- Refers to a nosebleed occurring without trauma or nose picking, ie. "Does it bleed on its' own?"

Both nostrils? - Only respond as No if the nose has only EVER bled from the left/right nostril. The answer is Yes if bleeding has occurred from both sides, either at the same time or at separate times.

After drug ingestion- Refers to nosebleeds occurring within seven days of taking aspirin, aspirin-containing preparations or other anti-inflammatory medication.

Cessation after compression- Refers to a nose bleed which stops with local pressure, not requiring consultation with a medical professional.

## Page 3

### Cutaneous symptoms

Bruise = Black and blue discolouration of the skin that is non-palpable (not raised)

Petechiae = “Pin-prick” bruising, usually red/purple in colour.

Hematoma = Bruising that is palpable/raised.

Definition of significant bruising/petechiae- Spontaneous bruises larger than 1 cm in diameter (the size of a pea) OR considered disproportionate to trauma by the investigator. All petechiae are considered to be significant.

Location of lesions- Exposed sites refers to the arms and legs. This is distinct from unexposed sites which would include the chest, back or abdomen.

Minimal or no trauma- Report Yes if the subject describes bruising which appears without injury or with injury that would not be expected to cause bruising.

A score for “Consultation” is recorded if the subject has ever sought medical attention or mentioned bruising to a medical professional.

### Bleeding from minor wounds

Minor wound = Superficial cut to the skin, such as that caused by a knife or scissors.

Definition of significant bleeding from minor wounds- Prolonged bleeding, lasting longer than 5 minutes, caused by a superficial cut.

A score for "Consultation" is recorded if the subject has ever sought medical attention for bleeding from a minor wound. This would include application of steri-strips.

Surgical hemostasis- Refers to stitching/suturing of a wound.

## Page 4

### Oral cavity bleeding

Oral cavity bleeding = Bleeding in the mouth.

Hemorrhagic bullae = Wet blood blisters.

Definition of significant oral cavity bleeding- Either spontaneous gum bleeding lasting for longer than a minute OR bites to lips/cheeks/tongue lasting longer than 5 minutes OR bleeding at tooth eruption requiring assistance by a physician.

Tooth eruption- Includes loss of teeth due to trauma.

### Tooth extraction

This section refers to extraction of teeth by a dentist and not to traumatic loss of teeth (which is included in the oral cavity bleeding category).

Definition of significant bleeding following tooth extraction- Any bleeding occurring after leaving the dentist's office or prolonged bleeding causing a delay in discharge from the dentist's office.

The total number of teeth extracted must be recorded, even in subjects not reporting problems with bleeding after extraction, as no bleeding on 2 or more occasions will generate a negative score.

For the positive control group, if the subject answers No to having had problems with bleeding after tooth extraction, remember to enquire about prophylaxis as this will generate a positive score.

It is most important to distinguish between extraction of deciduous and permanent teeth as extraction of permanent teeth would be expected to result in a higher risk of bleeding.

## Page 5

### GI bleeding

GI bleeding = Bleeding from esophagus, stomach or bowel.

Hematemesis = Vomiting fresh (red) or altered (coffee-ground) blood.

Melena = Passage of black, tarry stool.

Hematochezia = Passage of fresh blood per rectum.

Gastritis/ Ulcer = Gastritis refers to inflammation of the gastric mucosa (stomach lining). An ulcer refers to a break in the gastric mucosa. Both may cause abdominal pain, heartburn, hematemesis and/or melena.

Colitis = Inflammation of the colon, which can be due to inflammatory bowel disease, (eg. ulcerative colitis), ischaemia or infection. Symptoms include abdominal pain, fever and hematochezia.

Mallory-Weiss tear = A tear in the lining of the esophagus, at its' junction with the stomach. This usually occurs as a result of severe retching or vomiting and can cause hematemesis and/or melena.

Vascular malformation = In the gastrointestinal tract this usually refers to angiodysplasia whereby fragile mucosal blood vessels may bleed causing melena or hematochezia.

All GI bleeding is considered to be significant.

Bleeding associated with endoscopy should also be reported. How this is done will depend on why the endoscopy was performed.

- If the scope was indicated because of bleeding, the type of bleeding will be scored according to the categories on the questionnaire (ie: hematemesis, melena, etc) and then any identified pathology will also be indicated (ie: ulcer, colitis, etc). This will not change if the bleeding worsened after the scope, but in the "Other" space, the comment "worsened after endoscopy" will be written.

- If the scope was not being done because of bleeding, but because of abdominal pain for example, and the scope itself precipitated bleeding, then the type of bleeding will be indicated according to the categories on the questionnaire (ie: hematemesis, melena, etc). Again, any pathology identified during the scope will also be indicated (ie: ulcer, colitis, etc) and the "Other" category will be checked and the comment "precipitated by endoscopy" will be written.

## **Surgical bleeding**

The category of surgical bleeding does not include post-partum bleeding or bleeding after circumcision.

Definition of significant surgical bleeding- Any bleeding stated as abnormally prolonged by the surgeon OR causing a delay in discharge OR requiring some supportive treatment.

The total number of surgeries must be recorded, even in subjects not reporting problems with post-surgical bleeding, as no bleeding on 2 or more occasions will generate a negative score.

For the positive control group, if the subject answers No to having had problems with post-surgical bleeding, remember to enquire about prophylaxis as this generate a positive score.

**Menorrhagia**

Menorrhagia = Heavy periods.

Even if the subject does not report having a problem with menorrhagia, the questions regarding duration of menstruation and frequency of changing pads/tampons should always be asked as this symptom is often under-reported.

When asked which type of feminine product is used state the absorbency (ie. regular, super, super plus, etc.) and if both pads and tampons are used at the same time.

Add further details in the comments section if felt to be relevant, eg. flooding, passage of clots, changing pads/tampons during the night, marking the sheets at night, using diapers, etc.

Age of maximum severity- If there has been no change in severity over time then tick all of the boxes since menarche.



**Post-partum hemorrhage**

Post-partum hemorrhage = Vaginal bleeding occurring within 6 weeks of delivery of a baby.

Assisted delivery = The use of forceps or vacuum extraction to assist vaginal delivery.

Definition of significant post-partum hemorrhage- Any bleeding stated as abnormally prolonged by the obstetrician OR causing a delay in discharge OR requiring some supportive treatment.

**Muscle hematomas**

Muscle hematoma = A collection of blood within a muscle resulting in a functional component, eg. a calf muscle bleed with a limp. This would need to have been diagnosed by a doctor, except in someone with a severe bleeding problem who may have had the symptom previously.

All muscle hematomas are considered to be significant.

## **Hemarthrosis**

Hemarthrosis = A bleed into a joint. An ankle, knee or elbow bleed would be expected to cause swelling, pain and limitation of motion at the joint. A hip or shoulder bleed would need to have been confirmed by imaging studies.

All episodes of hemarthrosis are considered to be significant.

## **CNS bleeding**

CNS bleeding = Bleeding within the skull.

Subdural hemorrhage = Bleeding which occurs between the dura and the arachnoid.

Intracerebral hemorrhage = Bleeding which occurs within the brain tissue.

All episodes of CNS bleeding are considered to be significant.

**Other bleeding**

Cephalohematoma = Bleeding between the skull and the periosteum in a newborn baby. This usually occurs following delivery by vacuum extraction or forceps. It causes a soft swelling with distinct borders on the baby's head.

Venipuncture bleeding = Bleeding occurring after taking a venous blood sample.

Suction bleeding = Blood seen on suctioning of a baby's airway (via mouth or nostrils) immediately after birth.

Macroscopic hematuria = Blood in the urine that is visible to the naked eye.

All episodes of any of the above are considered to be significant.

If more than one of these events has occurred, the most severe presentation for each is summated to give the total score for this category. This is the **ONLY** category in which the scores are summated for individual bleeding episodes- in the other categories of bleeding symptoms, only the most severe presentation is recorded.