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No. 163-Gynaecological and Obstetric Management of Women With Inherited Bleeding Disorders

This document is based on a summary of the workshop entitled "Gynaecological and Obstetrical Management of Women with von Willebrand Disease" as presented at the First Canadian State-of-the-Art Conference on von Willebrand Disease, held in Montréal on May 8 and 9, 2003, to mark the 50th Anniversary of the Canadian Hemophilia Society.
This consensus document has been developed by a multidisciplinary committee consisting of an anesthesiologist, 2 hematologists, and an obstetrician/gynaecologist and has been endorsed by their relevant specialty bodies. It has been prepared with the express purpose of providing guidelines for both women with inherited bleeding disorders and for their caregivers regarding the gynaecological and obstetric management of these women, including appropriate anesthesia support where indicated.

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The text in part has been published by the Canadian Hemophilia Society in a document entitled "The Management of Women with Bleeding Disorders," prepared by the Subcommittee on Women with Bleeding Disorders of the Association of Hemophilia Clinic Directors of Canada. Members of this subcommittee include Michele David, MD, Montréal, QC; Christine Demers, MD, Québec City, QC; Diane Francoeur, MD, Montréal, QC; Bernadette Garvey, MD, Toronto, ON; Sara Israels, MD, Winnipeg, MB; David Lillicrap, MD, Kingston, ON; Georges-Etienne Rivard, MD, Montréal, QC; Mary Frances Scully, MD, St. John's, NL; Linda Vickars, MD, Vancouver, BC.
Key Words: Menorrhagia, Coagulation disorder, Pregnancy, Von Willebrand

Abstract

Objective: The prevalence of bleeding disorders, notably von Willebrand disease (vWD), among adult women with objectively documented menorrhagia is consistently reported to be 10% to 20% and is even higher in adolescents presenting with menorrhagia.

Options: Diagnostic tools and specific medical and, where appropriate, surgical alternatives to management are reviewed and evidence-based recommendations presented.

Evidence: A MEDLINE search of the English literature between January 1975 and November 2003 was performed using the following key words: menorrhagia, uterine bleeding, pregnancy, von Willebrand, congenital bleeding disorder, desmopressin/DDAVP, tranexamic acid, oral contraceptives,

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medroxyprogesterone, therapy, hysterectomy, anesthesia, epidural, spinal. Recommendations from other society guidelines were reviewed.

Values: The quality of evidence reported in this document has been described USing the Evaluation of Evidence criteria outlined in the Report of the Canadian Task Force on the Preventive Health Exam (Table 1).[13]

Recommendations:

1. Inherited bleeding disorders should be considered in the differential diagnosis of all patients presenting with menorrhagia (II-2B). The graphical scoring system presented is a validated tool which offers a simple yet practical method that can be used by patients to quantify their blood loss (II-2B).
2. Because underlying bleeding disorders are frequent in women with menorrhagia, physicians should consider performing a hemoglobin/hematocrit, platelet count, ferritin, PT (INR) and APTT in women with menorrhagia. In women who have a personal history of other bleeding or a family history of bleeding, further investigation should be considered, including a vWD workup (factor VIII, vWF antigen, and vWF functional assay) (II-2B).
3. Treatment of menorrhagia in women with inherited bleeding disorders should be individualized (III-B).
4. An inherited bleeding disorder is not a contraindication to hormonal therapy (oral contraceptives [II-IB], depot medroxyprogesterone acetate (DMPA) [II-3B], danazol [II-2B], GnRH analogs [II-3B]) or local treatments (levonorgestrel-releasing IUS [II-IB]) and non-hormonal therapy (antifibrinolytic drug tranexamic acid [II-IB]) as well as desmopressin (II-IB). These therapies represent first line treatment. Blood products should not be used for women with mild bleeding disorders (III-A).
5. In women who no longer want to preserve their fertility, conservative surgical therapy (ablation) and hysterectomy may be options (III-B). Clinicians may consult the "SOGC Clinical Practice Guideline: Guidelines for the Management of Abnormal Uterine Bleeding" for an in-depth discussion of the available therapeutic modalities, both medical and surgical. To minimize the risk of intraoperative and post-operative hemorrhage, coagulation factors should be corrected preoperatively with post-operative monitoring (II-IB).
6. Girls growing up in families with a history of vWD or other inherited bleeding disorders should be tested premenarchally to determine whether or not they have inherited the disease to allow both the patient and her family to prepare for her first and subsequent menstrual periods (III-C).
7. In adolescents presenting with menorrhagia, an inherited bleeding disorder should be excluded (III-B). When possible, investigation should be undertaken before oral contraceptive therapy is instituted, as the hormonally induced increase in factor VIII and vWF may mask the diagnosis (II-B).
8. Pregnancy in women with inherited bleeding disorders may require a multidisciplinary approach. A copy of their recommendations should be given to the patient and she should be instructed to present it to the health care provider admitting her to the birthing centre. Women with severe bleeding disorders or with a fetus at risk for a severe bleeding disorder should deliver in a hospital (level three) or where there is access to consultants in obstetrics, anesthesiology, hematology, and pediatrics (III-C).
9. Vacuum extraction, forceps, fetal scalp electrodes, and fetal scalp blood sampling should be avoided if the fetus is known or thought to be at risk for a congenital bleeding disorder. A Caesarean section should be performed for obstetrical Indications only (II-2C).
10. Epidural and spinal anesthesia are contraindicated if there is a coagulation defect. There is no contraindication to regional anesthesia if coagulation is normalized. The decision to use regional anesthesia should be made on an individual basis (III-C).
11. The risk of early and late postpartum hemorrhage is increased in women with bleeding disorders. Women with inherited bleeding disorders should be advised aboutthe possibility of excessive postpartum bleeding and instructed to report this immediately (III-B).
12. Intramuscular injections, surgery, and circumcision should be avoided in neonates at risk for a severe hereditary bleeding disorder until adequate workup/preparation are possible (III-B).