



NATIONAL HEMOPHILIA FOUNDATION
for all bleeding and clotting disorders

MASAC Document #197
(Replaces #185)

**MASAC RECOMMENDATIONS REGARDING GIRLS AND WOMEN WITH
INHERITED BLEEDING DISORDERS**

The following recommendations were approved by the Medical and Scientific Advisory Council (MASAC) of the National Hemophilia Foundation on April 18, 2010, and adopted by the NHF Board of Directors on June 26, 2010.

Inherited bleeding disorders are under-recognized as causes of bleeding in girls and women. However, recent data have documented that 50% of girls and women who are carriers for hemophilia A or B have factor VIII or IX levels below 50%, putting them in the category of mild hemophilia and causing them to have an increased risk of bleeding, especially during menstruation and at the time of an accident or surgery. Moreover, 1%-2% of the general population may have laboratory evidence of von Willebrand disease. Thus 1%-2% of all girls and women may have inherited this bleeding tendency and may also have heavy bleeding during menstruation or surgery.

With this information in mind, MASAC recommends the following:

A. Management

1. Von Willebrand disease (VWD) and other inherited bleeding disorders should be considered in the differential diagnosis of all girls and women presenting with menorrhagia, and an appropriate workup should be undertaken. Initial testing should include a CBC, PT, PTT, and a TT or fibrinogen. Additional testing specifically for VWD should include factor VIII activity, ristocetin cofactor, von Willebrand antigen, and von Willebrand multimers. This workup should be done in consultation with a hematologist who is well versed in the diagnosis of inherited bleeding disorders.
2. The above laboratory evaluation should be considered in all girls and women scheduled for endometrial ablation and/or hysterectomy for menorrhagia or dysfunctional uterine bleeding who also have mucous membrane bleeding and/or a family history of bleeding.
3. If there is a positive family history of a bleeding disorder, girls and women should have the appropriate factor activity level determined as soon as feasible and definitely prior to any planned surgical procedure regardless of age.
4. For bleeding, trauma, and before and after any surgical procedure, females with a factor VIII level less than 50% may be treated with recombinant factor VIII or DDAVP (if DDAVP challenge has shown her to be responsive).

5. If a female with a factor IX level below 50% has a clinically significant bleeding episode, she may be treated with recombinant factor IX.
6. For clinical bleeding episodes, females with abnormal von Willebrand studies may be treated with DDAVP or a virally inactivated factor VIII product that contains von Willebrand factor (Alphanate, Humate P, Koate DVI, Wilate). Alphanate, Humate P, and Wilate are approved by the FDA for treatment of VWD. (See MASAC Document #190, *MASAC Recommendations Regarding the Treatment of von Willebrand Disease*, for specific details.)
7. Cryoprecipitate and fresh frozen plasma should not be used unless the patient is at risk of life-threatening bleeding and a Factor VIII/von Willebrand Factor concentrate is not rapidly available. (See MASAC Document #195, *MASAC Recommendations Concerning the Treatment of Hemophilia and Related Bleeding Disorders*, Revised April 2010).

B. Outreach, Education, and Research

1. Much progress has been made in increasing awareness of girls and women's bleeding disorders by both the general public and clinicians who are primary care providers for girls and women. Nonetheless, the national outreach and education program should be continued. The target audiences should be health care professionals (e.g. pediatricians, hematologists/oncologists, internists, OB/GYN, family practitioners, and dentists as well as nurse practitioners in these fields), women's health advocates, and the general public.
2. NHF should continue to work with NHLBI and CDC to develop a national research agenda on women's bleeding disorders.

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