



Summary of the Guidelines on the Management of von Willebrand Disease (VWD)

(a collaborative effort of ASH ISTH NHF WFH)

1 In patients with VWD with a history of severe and frequent bleeds, routine treatment to prevent bleeds (*prophylaxis*) rather than no prophylaxis should be considered.

2a In patients who might get treated with desmopressin (*i.e. DDAVP or Stimate*) and who have a baseline von Willebrand factor (VWF) level of less than 30%, a test of whether desmopressin will work for the patient (*trial*) should be performed.

2b In patients who have a VWF level of less than 30%, desmopressin should not be used for treatment until the results of the desmopressin trial are available.

3 In patients with VWD and cardiovascular disease, who need treatment with antiplatelet agents or anticoagulant therapy (*both prevent clotting*), the necessary medication should be given. Additional treatment with factor concentrate or desmopressin might be needed.

4a After major surgery, factor VIII (FVIII) and VWF activity levels should be kept at greater than or equal to 50% for at least 3 days.

4b After major surgery, VWF and FVIII should both be targeted to have an activity level of greater than or equal to 50% for at least 3 days.

5a In patients undergoing minor surgery or minor invasive tests, VWF activity levels should be increased to greater than or equal to 50% with a combination of tranexamic acid (*i.e. Lysteda*) with either desmopressin or factor concentrate.

5b Tranexamic acid should be used in patients with type 1 VWD with baseline VWF activity levels of greater than 30% and mild bleeding symptoms undergoing minor mucosal (*lining of the internal organs*) procedures (*i.e. colonoscopy*).

6a In people with VWD and heavy menstrual bleeding, who do not wish to become pregnant, hormonal therapy (*i.e. birth control pill or hormonal IUD like Mirena*) or tranexamic acid should be given rather than desmopressin.

6b In people with VWD and heavy menstrual bleeding, who wish to become pregnant, tranexamic acid should be given rather than desmopressin.

7 In pregnant people with VWD, who wish to receive spinal, epidural, or combined spinal-epidural pain management options during labor, a VWF activity level of 50% - 150% should be targeted.

8 In pregnant people with type 1 VWD or low VWF levels in any other type of VWD, tranexamic acid should be used after the delivery of a baby (*post-partum period*).

To read the VWD Guidelines in full, please go to:

<https://ashpublications.org/bloodadvances/article/5/1/301/474884/ASH-ISTH-NHF-WFH-2021-guidelines-on-the-management>