

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>