Appendix B: Inherited Bleeding Disorders Summary

Inherited Bleeding Disorders	Possible Hemostatic Treatments	Other information
Factor VIII (8) Deficiency (Hemophilia A) *Classifications: Severe, Moderate, Mild	• Factor VIII (8) • Desmopressin (DDAVP) • Tranexamic Acid *Desmopressin not indicated or ineffective for severe Hemophilia A	Normal FVIII (8) levels 50 - 150 % (reported as 0.50 - 1.50 units)
Factor IX (9) Deficiency (Hemophilia B or Christmas Disease) *Classifications: Severe, Moderate, Mild	• Factor IX (9) • Tranexamic Acid *Desmopressin not indicated for Hemophilia B	Normal FIX (9) levels 50 – 150 % (reported as 0.50 - 1.50 units)
von Willebrand Disease (vWD) • Sub type 1 and 1C (Decreased amount of vWF) • Sub types 2A, 2B, 2N, 2M (vWF function disorder) • Sub type 3 (No vWF production)	• vWF/FVIII (8) concentrate. • Desmopressin (DDAVP) • Tranexamic Acid *Desmopressin not indicated for type 3 or type 2B with risk of thrombocytopenia	Normal von Willebrand levels • VWF.Ricof: 0.50 - 1.50 units (Activity level = vWF function) • vWF:Ag: greater than 1.50 units (Antigen level = amount of vWF)
Factor XI (11) Deficiency (Old term: Hemophilia C)	• Factor XI (11)	Variable bleeding symptoms that do not correlate with FXI (11) level therefore treatment (if indicated) is individualized
Platelet disorders	• Platelet transfusion • Desmopressin (DDAVP) • Tranexamic Acid	
Rare factor deficiencies [e.g.: fibrinogen, FII (2), FV (5), FVII (7), FX (10), FXIII(13)]	Various treatments	Treatment with factor replacement does not necessarily need to achieve laboratory reported 'normal range' and is individualized
Bleeding Disorder of Unknown Cause (BDUC) or Undefined Bleeding Disorder	• Desmopressin (DDAVP) • Tranexamic Acid	Defined as a person with a clear bleeding tendency (positive bleeding score) in the presence of normal hemostatic lab tests.
Ehler-Danlos Syndrome (Collagen Disorder)	• Desmopressin (DDAVP) • Tranexamic Acid • Use caution with fragile tissues	Refer to: https://www.ehlers-danlos.com/

This material has been prepared solely for use at Providence Health Care (PHC). PHC accepts no responsibility for use of this material by any person or organization not associated with PHC. A printed copy of this document may not reflect the current electronic version.