

Von Willebrand Disease (VWD): Management

Inherited: VWD is the most common of the inherited bleeding disorders, with a prevalence of approximately 1 percent when random laboratory screening is performed. It can be inherited or acquired. Inherited VWD has been classified into three types: 1, 2 (four subtypes), and 3.

Type	Inheritance	VWF activity (Ritocerin cofactor)	RIPA (Ritocerin-induced platelet aggregation)	Multimer pattern
Type 1	Autosomal dominant	decreased	decreased	Decreased large
Type 2A Type 2B Type 2M Type 2N	Autosomal dominant or recessive Autosomal dominant Autosomal dominant or recessive Autosomal recessive	Decreased Decreased Decreased Normal	Decreased Increased Decreased Normal	Decreased large Decreased large Uniform decreased Normal
Type 3 (severe)	Autosomal recessive	Markedly decreased or absent	Markedly decreased or absent	Undetectable

Acquired: VWD can develop in association with a number of disease states, including lymphoid malignancies, lymphoproliferative and myeloproliferative disorders, autoimmune disorders, conditions of high vascular flow, and some medications (eg, valproic acid)

Rx: The five categories of medications for the treatment of von Willebrand disease (VWD) include **desmopressin (DDAVP)**; replacement therapy with **von Willebrand factor (VWF)-containing concentrates**; **antifibrinolytic drugs**; topical therapy with **thrombin or fibrin sealant**; and **estrogen therapy** in some settings in women

Rx	Dose	Comments
DDAVP	0.3 mcg/kg over 20 min IV Nasal spray 1 spray/nostril	Type 1, +/- 2 not for type 3
VWF concentrates	20-60 u/Kg	requires clinical experience
Antifibrinolytic: Aminocaproic acid TRX acid	20-50 mcg/kg PO 4 times/day 10 mcg/kg 3 times/day	Great in dental/mucosal bleeding
IVIG	1 g/kg daily x 2 days	When associated with immune disease

Question:

A patient with known type 1 Von Willebrand's disease presents for emergency appendectomy. Which of the following is the best initial approach to treatment?

- a. FFP
- b. PRBC
- c. Cryoprecipitate
- d. DDAVP

Answer: D - For the treatment of minor bleeding or minor surgery, we suggest the use of intravenous or intranasal DDAVP in patients who have shown a prior response to this agent. If bleeding is not adequately controlled, a von Willebrand factor (VWF) concentrate should be used