

- There are case reports of thrombosis (including myocardial infarction) after infusion of DDAVP. It should be used with caution in patients with a history or risk of cardiovascular disease.⁵⁵

Recommendation 5.6.1:

- For patients with mild or moderate hemophilia A and carriers of hemophilia A, the WFH recommends considering desmopressin (DDAVP) as an option for treatment.
- **REMARK:** The WFH recommends testing DDAVP prior to therapeutic use to evaluate the individual FVIII response. The decision to use DDAVP must be based on the patient's baseline FVIII activity, the increment achieved, and the duration of treatment required.
- **REMARK:** In general, the most common adverse events observed are tachycardia, flushing, tremor, abdominal discomfort, and headache, especially during rapid infusion, and are mostly mild and transient. However, hypotension and/or severe hyponatremia can also occur.
- **REMARK:** For pregnant women during labour and delivery, the WFH recommends caution in the use of DDAVP, and it should be avoided in pre-eclampsia and eclampsia.
- **REMARK:** With more than 3 consecutive days of dosing, the therapeutic response may decrease (tachyphylaxis) and the risk of complications rises; thus, clotting factor concentrates may be needed when higher factor levels are required for a prolonged period. **CB**

Recommendation 5.6.2:

- For adults, the WFH recommends DDAVP not be used for more than 3 consecutive days and only under close supervision. If DDAVP is administered twice in a single day, subsequent daily dosing should be limited to once per day.
- **REMARK:** In general, the most common adverse events observed are tachycardia, flushing, tremor, abdominal discomfort, and headache, especially during rapid infusion. However, hypotension and/or hyponatremia can also occur.
- **REMARK:** With more than 3 consecutive days of dosing, the therapeutic response may decrease (tachyphylaxis) and the risk of complications rises; thus, clotting factor concentrates may be needed when higher factor levels are required for a prolonged period. **CB**

Recommendation 5.6.3:

- For children, the WFH recommends using no more than 1 dose of DDAVP per day for no more than 3 consecutive days.
- **REMARK:** In general, the most common adverse events observed are tachycardia, flushing, tremor, abdominal discomfort, and headache, especially during rapid infusion. However, hypotension and/or hyponatremia can also occur.
- **REMARK:** With more than 3 consecutive days of dosing, the therapeutic response may decrease (tachyphylaxis) and the risk of complications rises; thus, clotting factor concentrates may be needed when higher factor levels are required for a prolonged period. **CB**

Recommendation 5.6.4:

- For children under 2 years of age, the WFH alerts that DDAVP is contraindicated due to increased risk of seizures as consequences of water retention and hyponatremia. **CB**

Recommendation 5.6.5:

- For patients at risk of cardiovascular disease or thrombosis, the WFH recommends that DDAVP should be used with caution due to the risk of thromboembolism and myocardial infarction. **CB**

Tranexamic acid

- Tranexamic acid is an antifibrinolytic agent that competitively inhibits the activation of plasminogen to plasmin. It promotes clot stability and is useful as adjunctive therapy for some types of hemophilic bleeding.⁶⁵
- Treatment with tranexamic acid alone is of no value in the prevention of hemarthroses in hemophilia.⁶⁵
- Tranexamic acid is useful for treating superficial soft tissue and mucosal bleeds (e.g., oral bleeding, epistaxis, and menorrhagia).⁶⁶⁻⁶⁸
- Tranexamic acid is particularly valuable in the setting of dental surgery and may be used to control oral bleeding associated with eruption or shedding of teeth.^{67,69}
- See also Chapter 2: Comprehensive Care of Hemophilia and Chapter 7: Treatment of Specific Hemorrhages.

Dosage/administration

- Tranexamic acid is usually given as oral tablets (25 mg/kg/dose) 3-4 times daily. It can also be given by intravenous infusion (10 mg/kg/dose) 2-3 times daily. It is also available as an oral rinse.
- Gastrointestinal upset (nausea, vomiting, or diarrhea) may rarely occur as a side effect of tranexamic acid, but these symptoms usually resolve if the dosage is reduced. When administered intravenously, tranexamic acid must be infused slowly as rapid injection may result in dizziness and hypotension.
- A syrup formulation of tranexamic acid is also available for pediatric use. If this is not obtainable, a tablet can be crushed finely and dissolved in clean water for topical use on bleeding mucosal lesions.
- Tranexamic acid is commonly prescribed for 7 days following dental extractions to prevent postoperative bleeding.
- Tranexamic acid is excreted by the kidneys, and the dose must be reduced if there is renal impairment in order to avoid toxic accumulation.
- The use of tranexamic acid is contraindicated for the treatment of hematuria as its use may prevent dissolution of clots in the ureter, leading to serious obstructive uropathy and potentially permanent loss of renal function.
- Tranexamic acid is also contraindicated in the setting of thoracic surgery where it may result in the development of insoluble hematomas.