PHARMACY FRIDAY PEARL | HEMATOLOGY | HEMOPHILIA

Review of Factor Products for Hemophilia

A Pharmacy Friday Pearl reviewing factor VIII, factor IX, recombinant factor VIIa, and activated prothrombin complex concentrate selection for hemophilia-related bleeding.

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Legacy source conversion · 6 min read · 14 references

Introduction

Hemophilia A is a factor VIII deficiency and hemophilia B is a factor IX deficiency. Both are lifelong bleeding disorders that may require urgent factor replacement when patients present with clinically significant bleeding.

Factor concentrates include plasma-derived and recombinant products. Products differ by purity, generation, cell line exposure, human or animal protein exposure, and half-life. Some recombinant products have extended half-life formulations that allow less frequent dosing.

Key Points

- For severe bleeding without inhibitors, the source Pearl lists factor VIII 50 units/kg for hemophilia A and factor IX 100-140 units/kg for hemophilia B to target 80-100% replacement.

- For life-threatening bleeding with inhibitors, recombinant factor VIIa 90 mcg/kg or activated prothrombin complex concentrate 75-100 units/kg are listed bypassing options.
- Factor VIII and factor IX products carry inhibitor and hypersensitivity considerations; bypassing agents carry thrombotic and drug-interaction concerns.
- Patients receiving emicizumab prophylaxis require extra caution with aPCC because of thrombosis and thrombotic microangiopathy risk.

Pharmacology and Dosing

TOPIC	PRACTICAL DETAIL
Factor VIII products	Dose range in the source table: 25-50 units/kg. Factor VIII replacement supports clot formation by participating with activated factor IX to activate factor X.
Factor IX products	Dose range in the source table: 50-140 units/kg. Factor IX replacement temporarily restores hemostasis in hemophilia B.
Recombinant factor VIIa	Dose listed: 90 mcg/kg/dose. Administer as IV bolus over 2-5 minutes; the source notes repeat dosing every 2 hours until hemostasis is achieved.
aPCC / FEIBA	Dose listed: 50-100 units/kg. Administer by IV injection or infusion at a maximum rate of 2 units/kg/minute and complete infusion within 3 hours.
Compatibility and warnings	The source lists normal saline for factor VIIa and flags interactions with emicizumab, antifibrinolytics, factor products, and PCC products depending on agent.

CLINICAL PEARL

Match the replacement or bypassing strategy to hemophilia type, bleeding severity, and inhibitor status. In hemophilia A patients receiving emicizumab prophylaxis, the source Pearl favors rFVIIa over FEIBA for acute bleeds or procedures.

Evidence Overview

SOURCE	DESIGN	PRACTICAL TAKEAWAY
Schneiderman 2004	Retrospective chart review of 5 hemophilia patients with inhibitors receiving sequential aPCC and rFVIIa.	No clinical or laboratory evidence of thrombosis, thrombocytopenia, or DIC was reported with careful inpatient monitoring.
Astermark 2007	Prospective open-label randomized crossover study of 66 hemophilia A patients and 96 bleeding episodes.	FEIBA and rFVIIa had similar overall efficacy at 6 hours, though some patients responded better to one product.
Young 2008	Randomized multicenter crossover double-blind study of hemophilia patients with inhibitors.	A single 270 mcg/kg rFVIIa dose was reported as safe and effective compared with repeated 90 mcg/kg dosing and aPCC.
Borg 2015	Non-interventional retrospective review of acquired hemophilia A patients treated with FEIBA.	Overall efficacy resolving bleeding episodes was 88%; thrombotic adverse effects occurred in 5 patients.
Olivieri 2020 / Yan 2020	Real-world cohort and chart analyses comparing recombinant factor VIII products.	Long-acting products supported reduced dosing frequency with comparable bleeding outcomes across factor VIII products.

BOTTOM LINE

Treat the bleed and inhibitor status first.

Patients with hemophilia A or B may be managed with standard-acting or long-acting products depending on the product, indication, and patient-specific factors.

Long-acting factor VIII products have comparable bleeding outcomes in the cited real-world evidence and may reduce dosing frequency.

For patients with inhibitors, bypassing agents such as recombinant factor VIIa or aPCC are key options, but drug interactions and thrombotic risk must be considered.

References

1. Centers for Disease Control and Prevention. Hemophilia treatment. Retrieved February 19, 2021.
2. UpToDate. Retrieved February 22, 2021.
3. Clinical Pharmacology. Retrieved February 22, 2021.
4. National Hemophilia Foundation. MASAC guidelines for emergency department management of individuals with hemophilia and other bleeding disorders.
5. National Hemophilia Foundation. Products licensed in the United States. Retrieved February 19, 2021.
6. Schneiderman J, Nugent DJ, Young G. Sequential therapy with activated prothrombin complex concentrate and recombinant factor VIIa. *Haemophilia*. 2004;10(4):347-351.
7. Astermark J, Donfield SM, DiMichele DM, et al. A randomized comparison of bypassing agents in hemophilia complicated by an inhibitor. *Blood*. 2007;109(2):546-551.
8. Young G, Shafer FE, Rojas P, Seremetis S. Single-dose rFVIIa vs standard rFVIIa and APCC for home treatment of joint bleeds. *Haemophilia*. 2008;14(2):287-294.
9. Borg JY, Negrier C, Durieu I, et al. FEIBA in acquired haemophilia A. *Haemophilia*. 2015;21(3):330-337.
10. Olivieri M, Sommerer P, Maro G, Yan S. Prophylactic use and clinical outcomes in hemophilia A. *Eur J Haematol*. 2020;104(4):310-317.
11. Yan S, Maro GS, Desai V, Simpson ML. Real-world FVIII product analysis. *J Manag Care Spec Pharm*. 2020;26(10):1258-1265.
12. Davis J, Yan S, Matsushita T, et al. Recombinant factor IX prophylaxis systematic review. *J Med Econ*. 2019;22(10):1014-1021.
13. Shapiro AD, Di Paola J, Cohen A, et al. Recombinant human blood coagulation factor IX in previously untreated patients. *Blood*. 2005;105(2):518-525.
14. Srivastava A, Santagostino E, Dougall A, et al. WFH Guidelines for the Management of Hemophilia, 3rd edition. *Haemophilia*. 2020;26 Suppl 6:1-158.

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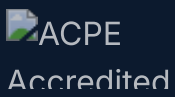

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