

Title: Hemophilia Agents

Policy #: Rx.01.149

Application of pharmacy policy is determined by benefits and contracts. Benefits may vary based on product line, group, or contract. Some medications may be subject to precertification, age, quantity, or formulary restrictions (ie limits on non-preferred drugs). Individual member benefits must be verified.

This pharmacy policy document describes the status of pharmaceutical information and/or technology at the time the document was developed. Since that time, new information relating to drug efficacy, interactions, contraindications, dosage, administration routes, safety, or FDA approval may have changed. This Pharmacy Policy will be regularly updated as scientific and medical literature becomes available. This information may include new FDA-approved indications, withdrawals, or other FDA alerts. This type of information is relevant not only when considering whether this policy should be updated, but also when applying it to current requests for coverage.

Members are advised to use participating pharmacies in order to receive the highest level of benefits.

Intent:

The intent of this policy is to communicate the medical necessity criteria for **Advate®**, **Adynovate®**, **Afstyla®**, **Alphanate®**, **Alphanine SD®**, **Alprolix®**, **Bebulin®**, **Benefix®**, **Coagadex®**, **Corifact®**, **Eloctate®**, **Esperoct®**, **Feiba NF®**, **Helixate FS®**, **Hemlibra®**, **Hemofil M®**, **Humate P®**, **Idelvion®**, **Ixinity®**, **Jivi®**, **Koate DVI®**, **Kogenate FS®**, **Kovaltry®**, **Monoclate P®**, **Mononine®**, **Novoeight®**, **Novoseven RT®**, **Nuwiq®**, **Obizur®**, **Profilnine®**, **Rebinyn®**, **Recombinate®**, **Riastap®**, **Rixubis®**, **Sevenfact®**, **Tretten®**, **Vonvendi®**, **Wilate®**, and **Xyntha®/Xyntha® Solofuse™** as provided under the member's prescription drug benefit.

Description:

Hemophilia is a bleeding disorder caused by clotting factor deficiencies. Hemophilia is a rare X-linked, congenitally acquired disease that leads to a deficiency in coagulation factor, VIII (hemophilia A) or IX (hemophilia B). Hemophilia A or B occurs in approximately 1 out of 10,000 births. Hemophilia A is more common than hemophilia B, representing approximately 80-85% of hemophilia cases. The disorder is further characterized by the percentage of serum clotting factor activity as compared to the normal range:

- **Severe hemophilia:** clotting factor levels are less than 1% of normal
- **Moderate hemophilia:** clotting factor levels are 1-5% of normal
- **Mild hemophilia:** clotting factors are 5-40% of normal

Treatment of hemophilia generally involves an infusion of the deficient factors to allow for more normalized clotting response and decreased incidence of uncontrolled bleeding events.

Treatment regimens are individualized based on disease severity and may include prophylactic infusions of clotting factors, on demand infusions of clotting factors, or a combination of both. Perioperative infusions are also part of the treatment regimen.

While Hemophilia A and hemophilia B are the most common types of hemophilia, 1 in every 500,000 to 2 million people have coagulation disorders caused by deficiencies in clotting factors other than factor VIII or IX. The products listed below include those with indications to treat these rare forms of coagulation disorders.

Riastap®	Fibrinogen concentrate, human					X			X		
Rixubis®	Factor IX, recombinant		X						X	X	X
Sevenfact®	Factor VIIa, recombinant	X	X								
Tretten®	Factor XIII A subunit, recombinant							X (A subunit)			X
Vonvendi®	Von Willebrand Factor Recombinant				X				X		
Wilate®	von Willebrand/ factor VIII complex, human				X				X		
Xyntha® [Solufuse]	Recombinant factor VIII	X							X	X	X

Policy:

Advate®, Adynovate®, Afstyla®, Alphanate®, Alphanine SD®, Alprolix®, Bebulin®, Benefix®, Coagadex®, Corifact®, Eloctate®, Esperoct®, Feiba NF®, Helixate FS®, Hemlibra®, Hemofil M®, Humate P®, Idelvion®, Ixinity®, Jivi®, Koate DVI®, Kogenate FS®, Kovaltry®, Monoclate P®, Mononine®, Novoeight®, Novoseven RT®, Nuwiiq®, Obizur®, Profilnine®, Rebinyn®, Recombinate®, Riastap®, Rixubis®, Sevefact®, Tretten®, Vonvendi®, Wilate®, Xyntha®/Xyntha® Solofuse are approved when there is a diagnosis of an FDA approved indication.

Approval duration: 2 years

Black Box Warning as shown in the drug Prescribing Information:

Feiba® NF

Thrombotic/Thromboembolic events: Thrombotic and thromboembolic events have been reported during postmarketing surveillance following infusion of anti-inhibitor coagulant complex, particularly following the administration of high doses and/or in patients with thrombotic risk factors. Monitor patients receiving anti-inhibitor coagulant complex for signs and symptoms of thromboembolic events.

Hemlibra®

Cases of thrombotic microangiopathy and thrombotic events were reported when on average a cumulative amount of >100 U/kg/24 hours of activated prothrombin complex concentrate was administered for 24 hours or more to patients receiving HEMLIBRA prophylaxis. Monitor for the development of thrombotic microangiopathy and thrombotic events if aPCC is administered. Discontinue aPCC and suspend dosing of HEMLIBRA if symptoms occur.

Novoseven®, Sevenfact®

Serious arterial and venous thrombotic events following administration of Novoseven and Sevenfact RT have been reported. Discuss the risks and explain the signs and symptoms of thrombotic and thromboembolic events to patients who

will receive Novoseven and Sevenfact RT. Monitor patients for signs and symptoms of activation of the coagulation system and for thrombosis.

Guidelines:

Refer to the specific manufacturer's prescribing information for administration and dosage details and any applicable Black Box warnings.

BENEFIT APPLICATION

Subject to the terms and conditions of the applicable benefit contract, the applicable drug(s) identified in this policy is (are) covered under the prescription drug benefits of the Company's products when the medical necessity criteria listed in this pharmacy policy are met. Any services that are experimental/investigational or cosmetic are benefit contract exclusions for all products of the Company.

References:

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Applicable Drugs:

Inclusion of a drug in this table does not imply coverage. Eligibility, benefits, limitations, exclusions, precertification/referral requirements, provider contracts, and Company policies apply.

Drug Names

Advate®

Adynovate®

Afstyla®

Alphanate®

Alphanine SD®

Alprolix®

Bebulin®

BeneFIX®

Coagadex®

Corifact®

Eloctate®

Esperoct®

Feiba NF®

Helixate FS®

Hemlibra®

Hemofil M®

Humate-P®

Idelvion®

Ixinity®

Jivi®

Koate-DVI®

Kogenate FS®

Kovaltry®

Monoclate-P®

Mononine®

Novoeight®

Novoseven RT®

Nuwiq®

Obizur®

Profilnine SD®

Rebinyn®

Recombinate®

Riastap®

Rixubis®

Sevenfact®

Tretten®

Vonvendi®

Wilate®

Xyntha®

Cross References:

Rx.01.33 Off-Label Use

Policy Version Number:	21.00
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