

PHARMACY POLICY STATEMENT Marketplace

DRUG NAME	Hemophilia and Other Clotting Disorders
BENEFIT TYPE	Medical
STATUS	Prior Authorization Required

Hemophilia is the most common severe hereditary hemorrhagic disorder. Both hemophilia A and B result from factor VIII and factor IX protein deficiency or dysfunction, respectively, and is characterized by prolonged and excessive bleeding after minor trauma or sometimes even spontaneously. Hemophilia A is more common than hemophilia B, representing 80–85% of the total hemophilia population.

Hemophilia and Other Clotting Disorders will be considered for coverage when the following criteria are met:

Hemophilia A (Factor VIII Deficiency)

For **initial** authorization:

- 1. Member has diagnosis of Hemophilia A (congenital Factor VIII deficiency); AND
- 2. For Jivi, member must be 12 years of age or older; AND
- 3. Medication is being prescribed by or in consultation with a hematologist; AND
- 4. Medication will be used for applicable situations listed in Table A or for Immune Tolerance Induction (ITI); AND
- 5. If request is for ITI, member must have severe hemophilia (factor level < 1%) with inhibitors (FVIII titre > 0.6 BU), and meet <u>one</u> of the following:
 - a. Inhibitor titre < 10 BU/mL or titre fails to fall below 10 BU/mL within a year;
 - b. Member is having severe or life-threatening bleeding;
 - c. Member is having frequent bleeding and is being considered for bypassing agent prophylaxis: AND
- 6. If request is for Altuviiio, member has had a trial and failure of a standard half-life FVIII product OR extended half-life FVIII product; AND
- 7. Member's recent weight (kg), history of bleeds, number of <u>as needed doses</u> on hand, and inhibitor status have been provided for review.
- **8. Dosage allowed:** Per package insert of individual drug. For ITI, dosages may range from 50 IU/kg three times weekly to 200 IU/kg daily depending on titer inhibitor levels.

If all the above requirements are met, the medication will be approved for 30 days for perioperative management or 6 months for all other cases.

Note: Approval will be for requested dosage, but no more than +/- 5-10% of prescribed assays. For patients on prophylaxis: the number of <u>as needed doses</u> the patient has on hand will be taken into consideration for treatment of acute bleeding episodes. A maximum of 5 <u>as needed doses</u> will be permitted at a time.



For **reauthorization**:

- 1. Member's recent weight (kg), history of bleeds, number of <u>as needed doses</u> on hand, and inhibitor status have been provided for review; AND
- 2. Member has experienced positive clinical response from the use of factor; AND
- 3. If request is for a dosage increase, provider must submit a clinical rationale supported by chart notes; AND
- 4. For ITI, chart notes have been provided to show both of the following:
 - Member continues to need ITI (e.g., inhibitor is detectable (> 0.6 BU), FVIII recovery < 66% of expected, FVIII half-life is < 7 hours); AND
 - b. Member has shown at least 20% decline in the inhibitor titre level since the previous approval.

If all the above requirements are met, the medication will be approved for an additional 6 months.

Hemophilia B (Factor IX Deficiency)

For **initial** authorization:

- 1. Member has diagnosis of Hemophilia B (congenital Factor IX deficiency); AND
- 2. For Ixnity, member must be 12 years of age or older; AND
- 3. For AlphaNine, member must be 17 years of age or older; AND
- 4. Medication is being prescribed by or in consultation with a hematologist; AND
- 5. Medication will be used for applicable situations listed in Table A or for Immune Tolerance Induction (ITI); AND
- 6. If request is for ITI, member must have inhibitors (FIX titre ≥ 0.3 BU) and prescriber must attest that benefit outweighs the risk of starting therapy; AND
- 7. Member's recent weight (kg), history of bleeds, number of <u>as needed doses</u> on hand, and inhibitor status have been provided for review.
- 8. Dosage allowed: Per package insert of individual drug.

If all the above requirements are met, the medication will be approved for 30 days for perioperative management or 6 months for all other cases.

Note: Approval will be for requested dosage, but no more than +/- 5-10% of prescribed assays. For patients on prophylaxis: the number of <u>as needed doses</u> the patient has on hand will be taken into consideration for treatment of acute bleeding episodes. A maximum of 5 <u>as needed doses</u> will be permitted at a time.

For **reauthorization**:

- 1. Member's recent weight (kg), history of bleeds, number of <u>as needed doses</u> on hand, and inhibitor status have been provided for review; AND
- 2. Member has experienced positive clinical response from the use of factor; AND
- 3. If request is for a dosage increase, provider must submit a clinical rationale supported by chart notes.



If all the above requirements are met, the medication will be approved for an additional 6 months.

FEIBA (anti-inhibitor coagulant complex)

For **initial** authorization:

- Member has a diagnosis of Hemophilia A or B with confirmed inhibitors (FVIII titre > 0.6 BU for hemophilia A or FIX titre ≥ 0.3 BU for hemophilia B); AND
- 2. Medication is being prescribed by or in consultation with a hematologist; AND
- 3. Medication will be used in one of the following situations:
 - a. On-demand treatment of acute bleeding episodes;
 - b. Perioperative management of bleeding;
 - c. Routine prophylaxis to prevent or reduce the frequency of bleeding episodes; AND
- 4. Member's recent weight (kg), history of bleeds, and inhibitor status have been provided for review; AND
- 5. If member is using Hemlibra, must have a clinical reason why a recombinant activated factor VII (rFVIIa) such as NovoSevenRT or Sevenfact cannot be used.
- 6. Dosage allowed: Per package insert.

If member meets all the requirements listed above, the medication will be approved for 30 days for perioperative management or 6 months for all other cases.

Note: Approval will be for requested dosage, but no more than +/- 5-10% of prescribed assays.

For **reauthorization**:

- 1. Member's recent weight (kg), history of bleeds, and inhibitor status have been provided for review; AND
- 2. Member has experienced positive clinical response from the use of factor; AND
- 3. If request is for a dosage increase, provider must submit a clinical rationale supported by chart notes.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 6 months.

Novoseven RT (Recombinant Factor VIIa)

For **initial** authorization:

- 1. Medication is being prescribed by or in consultation with a hematologist; AND
- 2. Medication is being used for the treatment of bleeding episodes OR perioperative management for one of the following diagnoses:
 - a. Hemophilia A or B with confirmed inhibitors (FVIII titre > 0.6 BU for hemophilia A or FIX titre ≥ 0.3 BU for hemophilia B);
 - b. Acquired hemophilia;
 - c. Congenital Factor VII (FVII) deficiency;



- d. Glanzmann's Thrombasthenia <u>and</u> platelet transfusion was either ineffective or contraindicated; AND
- 3. Member's recent weight (kg), history of bleeds, and inhibitor status (if applicable) have been provided for review.
- **4. Dosage allowed:** Per package insert.

If member meets all the requirements listed above, the medication will be approved for 30 days for perioperative management or 6 months for all other cases.

Note: Approval will be for requested dosage, but no more than +/- 5-10% of prescribed assays.

For **reauthorization**:

- 1. Member's recent weight (kg), history of bleeds, and inhibitor status (if applicable) have been provided for review; AND
- 2. Member has experienced positive clinical response from the use of factor; AND
- 3. If request is for a dosage increase, provider must submit a clinical rationale supported by chart notes.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 6 months.

Sevenfact (Recombinant Factor VIIa)

For **initial** authorization:

- 1. Member is 12 years of age or older; AND
- 2. Member has a diagnosis of Hemophilia A or B with confirmed inhibitors (FVIII titre > 0.6 BU for hemophilia A or FIX titre ≥ 0.3 BU for hemophilia B); AND
- 3. Medication is being prescribed by or in consultation with a hematologist; AND
- 4. Medication will be used as on-demand treatment of acute bleeding episodes; AND
- 5. Member's recent weight (kg), history of bleeds, and inhibitor status have been provided for review.
- 6. Dosage allowed: Per package insert.

If member meets all the requirements listed above, the medication will be approved for 6 months.

Note: Approval will be for requested dosage, but no more than +/- 5-10% of prescribed assays.

For **reauthorization**:

- 1. Member's recent weight (kg), history of bleeds, and inhibitor status have been provided for review; AND
- 2. Member has experienced positive clinical response from the use of factor; AND
- 3. If request is for a dosage increase, provider must submit a clinical rationale supported by chart notes.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 6 months.



Hemlibra (emicizumab-kxwh)

For **initial** authorization:

- 1. Member has diagnosis of Hemophilia A, with congenital factor VIII deficiency confirmed by blood coagulation testing; AND
- 2. Medication is being prescribed by or in consultation with a hematologist; AND
- 3. Member's recent weight (kg), history of bleeds, and inhibitor status have been provided for review; AND
- 4. For member with factor VIII inhibitors, member must meet the following:
 - a. Chart notes with documented positive test for inhibitors (titer > 0.6 BU/mL [Bethesda unit per milliliter]); OR
- 5. For member <u>without</u> factor VIII inhibitors, member must have severe hemophilia A (Factor VIII level <1%); AND
- 6. Bypassing agents (e.g., Feiba, NovoSeven RT, Sevenfact) are discontinued the day before starting Hemlibra (if applicable); AND
- 7. Prophylactic use of factor replacements are discontinued after loading dose period is finished.

Note: Factor VIII may be used as on-demand therapy for breakthrough bleeding.

8. Dosage allowed: 3 mg/kg subQ once weekly for the first 4 weeks, followed by a maintenance dose of 1.5 mg/kg once every week, OR 3 mg/kg once every 2 weeks, OR 6 mg/kg every 4 weeks.

If member meets all the requirements listed above, the medication will be approved for 6 months.

Note: Approval will be for the lowest number of vials to achieve requested dosage.

For **reauthorization**:

- 1. Member's recent weight in kilograms is documented on medication prior authorization request;
- 2. Chart notes have been provided showing that the member experienced a reduction in bleeding episodes compared to baseline.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 6 months.

Von Willebrand Disease (VWD)

For **initial** authorization:

- 1. Member has a diagnosis of Von Willebrand Disease (VWD); AND
- 2. For Vonvendi, member must be 18 years of age or older; AND
- 3. Medication is being prescribed by or in consultation with a hematologist; AND
- 4. Medication will be used for applicable situations listed in Table A; AND
- 5. Member has severe vWD (except Alphanate) OR Member has mild or moderate vWD and the use of desmopressin is known or suspected to be ineffective or contraindicated; AND
- 6. Member's recent weight (kg) and history of bleeds have been provided for review.



7. Dosage allowed: Per package insert of individual drug.

If member meets all the requirements listed above, the medication will be approved for 30 days for perioperative management, or 6 months for all other cases.

Note: Approval will be for requested dosage, but no more than +/- 5-10% of prescribed assays.

For **reauthorization**:

- 1. Member's recent weight (kg) and history of bleeds have been provided for review; AND
- 2. Member has experienced positive clinical response from the use of factor; AND
- 3. If request is for a dosage increase, provider must submit a clinical rationale supported by chart notes.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 6 months.

Miscellaneous Factors - (Obizur, Coagadex, Corifact, Tretten, Fibryga, RiaSTAP)

For initial authorization:

- 1. For Obizur, member must be 18 years of age or older with a baseline anti-porcine factor VIII inhibitor titer less than 20 BU; AND
- 2. Member has an FDA approved indication for use as listed in Table A; AND
- 3. Medication is being prescribed by or in consultation with a hematologist; AND
- 4. Member's recent weight (kg), history of bleeds, and fibrinogen level (if available, Fibryga and RiaSTAP only) have been provided for review.
- 5. Dosage allowed: Per package insert.

If member meets all the requirements listed above, the medication will be approved for 30 days for perioperative management or 6 months for all other cases.

Note: Approval will be for requested dosage, but no more than +/- 5-10% of prescribed assays.

For **reauthorization**:

- 1. Member's recent weight (kg) and history of bleeds have been provided for review; AND
- 2. Member has experienced positive clinical response from the use of factor; AND
- 3. If request is for a dosage increase, provider must submit a clinical rationale supported by chart notes.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 6 months.

Anti-Clotting Products - (ATryn, Ceprotin)



For **initial** authorization:

- 1. Member has an FDA approved indication for use as listed in Table A; AND
- 2. Medication is being prescribed by or in consultation with a hematologist; AND
- 3. Member's recent weight (kg) and chart notes supporting diagnosis have been provided for review.
- 4. Dosage allowed: Per package insert.

If member meets all the requirements listed above, the medication will be approved for 6 months.

For **reauthorization**:

- 1. Member's recent weight (kg) and documentation of positive clinical response have been submitted for review; AND
- 2. If request is for a dosage increase, provider must submit a clinical rationale supported by chart notes.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 6 months.

Table A

Drug Class	Drug Name	Indications
Recombinant Factor VIII (Hemophilia A)	Advate	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Afstyla	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Kovaltry	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Novoeight	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Nuwiq	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Recombinat e	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Xyntha	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
Extended Half-Life Recombinant Factor VIII (Hemophilia A)	Adynovate	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes



	Altuviiio	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Eloctate	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Esperoct	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Jivi	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
Plasma-Derived Factor	Hemofil M	Prevention and control of hemorrhagic episodes
VIII (Hemophilia A)	Koate	Prevention and control of bleeding episodes
Non-Factor (Hemophilia A)	Hemlibra	 Routine prophylaxis to prevent or reduce the frequency of bleeding episodes in adults and pediatric patients with hemophilia A with or without factor VIII inhibitors
	Benefix	 Hemophilia B (congenital factor IX deficiency) for: On-demand treatment and control of bleeding episodes Perioperative management of bleeding Routine prophylaxis to reduce the frequency of bleeding episodes
Recombinant Factor IX (Hemophilia B)	lxinity	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Rixubis	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Alprolix	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
Extended Half-Life Recombinant Factor IX (Hemophilia B)	Idelvion	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Rebinyn	 On-demand treatment and control of bleeding episodes Perioperative management Prevention and control of bleeding episodes
Plasma-Derived Factor IX (Hemophilia B)	AlphaNine SD	Prevention and control of bleeding episodes
Factor IX Complex (Hemophilia B)	Profilnine SD	Prevention and control of bleeding episodes



		Control and prevention of bleeding in patients with hemophilia A
von Willebrand Factor/Coagulation Factor VIII Complex	Alphanate	 Surgical and/or invasive procedures in adult and pediatric patients with von Willebrand Disease in whom desmopressin (DDAVP) is either ineffective or contraindicated. Not indicated for patients with severe VWD (Type 3) undergoing major surgery
		Hemophilia A
		 Treatment and prevention of bleeding in adults
	Humate-P	Von Willebrand disease
		 Treatment of spontaneous and trauma-induced bleeding episodes Perioperative management
(Human)		Children and adults with von Willebrand disease for:
	Wilate	 On-demand treatment and control of bleeding episodes Perioperative management
		Routine prophylaxis to reduce the frequency of bleeding episodes
		Adolescents and adults with hemophilia A for:
		On-demand treatment and control of bleeding episodes
		Routine prophylaxis to reduce the frequency of bleeding episodes
	Vonvendi	Adults with von Willebrand disease for:
vonWillebrand		 On-demand treatment and control of bleeding episodes Perioperative management
Recombinant Factor		Routine prophylaxis to reduce the frequency of bleeding episodes in patients with severe Type 3 von Willebrand disease receiving ondemand therapy.
		Hemophilia A and B with inhibitors for:
	Feiba	 On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
		Treatment of bleeding episodes and peri-operative management in
Bypassing Agent	NovoSeven RT	 adults and children with hemophilia A or B with inhibitors Congenital Factor VII (FVII) deficiency
		Glanzmann's thrombasthenia with refractoriness to platelet
		 transfusions, with or without antibodies to platelets Treatment of bleeding episodes and peri-operative management in
		adults with acquired hemophilia
	SevenFact	On-demand treatment of bleeding episodes in adults and adolescents with hemophilia A or B with inhibitors
Miscellaneous Factor	Obizur	On-demand treatment of bleeding episodes in adults with acquired hemophilia A
		Hereditary Factor X deficiency for:
	Coagadex	 Routine prophylaxis to reduce the frequency of bleeding episodes On-demand treatment and control of bleeding episodes



		 Perioperative management of bleeding in patients with mild, moderate and severe hereditary Factor X deficiency
	Corifact	 Routine prophylactic treatment and peri-operative management of surgical bleeding in patients with congenital Factor XIII deficiency
	Tretten	 Prophylaxis of bleeding in patients with congenital Factor XIII A- Subunit deficiency
	Fibryga	 Treatment of acute bleeding episodes in adults and children with congenital fibrinogen deficiency, including afibrinogenemia and hypofibrinogenemia
	RiaSTAP	 Treatment of acute bleeding episodes in adults and children with congenital fibrinogen deficiency, including afibrinogenemia and hypofibrinogenemia
Antithrombin	ATryn	Prevention of peri-operative and peri-partum thromboembolic events in patients with hereditary antithrombin deficiency
Protein C Concentrate	Ceprotin	 Treatment and prevention of venous thrombosis and purpura fulminans in patients with severe congenital Protein C deficiency

CareSource considers Hemophilia and Other Clotting Disorders not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
12/15/2016	Policy issued.
06/12/2018	Policy placed in a new format. Initial authorization length increased to 6 months.
10/05/2018	New drug Jivi added to the list of antihemophilic agents.
08/06/2019	New drug Esperoct added to the list of antihemophilic agents.
10/19/2019	Policy updated to include Hemlibra criteria.
08/01/2020	Hemlibra criteria updated to include hematologist. Requirement changed for members without Factor VIII inhibitors to align better with current practice and clinical trials.
04/02/2021	Title updated to encompass all bleeding disorder products. Table A created for all products, indications, and J codes. Added separate criteria set for hemophilia A, hemophilia B, Feiba, NovoSevenRT, Sevenfact, Von Willebrand Disease, miscellaneous factors, and anti-clotting products (previously only had one set of criteria for hemophilia factor replacement). Updated Hemlibra's weight requirement, reauth criteria, and dosage allowed section. Added approval instruction note for the factors and Hemlibra. Updated initial approval duration for all agents.



09/13/2022	Annual Review. Transferred to new template. Updated references. Removed discontinued medications from policy (Helixate, Kogenate). Updated Table A indications (VonVendi). Added baseline titer requirements for Obizur.
04/10/2023	Added Altuviiio and as needed acute bleed dosing guidance for prophylaxis to hemophilia A. Changed name from bleeding disorder agents to hemophilia and other clotting disorders. Added trial of Jivi (for extended half-life products) and Advate (for standard half-life products) for hemophilia A. Added a note that Hemlibra is preferred for long-term prophylaxis for hemophilia A. Removed trial of factor products, clinical reason factors cannot be used or poor venous access for patients who are not using factor products with Hemlibra.
01/05/2024	Added severe indication for perioperative management of bleeding for Coagadex; added indication of routine prophylaxis to reduce the frequency of bleeding episodes for Wilate; updated references
05/15/2024	Removed age limit for Ixinity.
07/02/2024	Removed Jivi and Advate trials and added a trial of an extended half-life or standard half-life FVIII product for Altuviiio.

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