

PHARMACY POLICY STATEMENT

Marketplace

DRUG NAME	Hemophilia and Other Clotting Disorders
BENEFIT TYPE	Pharmacy or medical: Alhemo, Hemlibra, Hymoviz, Qfitia Medical only: all others
STATUS	Prior Authorization Required

Hemophilia is the most common severe hereditary hemorrhagic disorder. Hemophilia A and B result from factor VIII and factor IX protein deficiency or dysfunction, respectively, and can result in prolonged and excessive bleeding after minor trauma. Spontaneous bleeding is also possible. 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 7 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 one 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 Altuviiro, 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 as needed doses 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 as needed doses the patient has on hand will be taken into consideration for treatment of acute bleeding episodes. A maximum of 5 as needed doses will be permitted at a time.

For **reauthorization**:

1. Member's recent weight (kg), history of bleeds, number of as needed doses 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:
 - a. 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 Ixniy, 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 as needed doses 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 $\pm 5\text{-}10\%$ of prescribed assays. For patients on prophylaxis: the number of as needed doses the patient has on hand will be taken into consideration for treatment of acute bleeding episodes. A maximum of 5 as needed doses will be permitted at a time.

For **reauthorization**:

1. Member's recent weight (kg), history of bleeds, number of as needed doses 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:

1. 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 and 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 without 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 3mg/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; AND
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.

Hypavzi (marstacimab-hncq)

For initial authorization:

1. Member is at least 12 years of age; AND
2. Medication is being prescribed by or in consultation with a hematologist; AND
3. Member has diagnosis of one of the following:
 - a. Severe hemophilia A (indicated by 1% or less of normal circulating factor VIII) or
 - b. Moderate to severe hemophilia B (indicated by 2% or less of normal circulating factor IX); AND
4. Member does NOT have factor inhibitors; AND
5. Member has tried and failed a compliant regimen of factor prophylaxis or Hemlibra; AND
6. Prophylactic therapy will be discontinued before initiating Hypavzi; AND
7. Member will NOT use Hypavzi for breakthrough bleeds.
8. **Dosage allowed/Quantity limit:** 300 mg subcutaneous loading dose, followed by 150 mg subcutaneous once weekly dose. Consider dose adjustment to 300 mg once weekly in patients weighing more than 50 kg when control of bleeding events is judged to be inadequate. Quantity limit: 4 syringes/pens (150 mg/mL) per 28 days after loading dose

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

For reauthorization:

1. Chart notes must show 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.

Alhemo (concizumab-mtci)

For **initial** authorization:

1. Member is at least 12 years of age; AND
2. Medication is being prescribed by or in consultation with a hematologist; AND
3. Member has a documented diagnosis of congenital hemophilia A or B; AND
4. For members with inhibitors, member meets **BOTH** of the following:
 - a. Documented history of factor inhibitor, equal to or above 0.6 Bethesda Units (BU);
 - b. Prescribed, or in need of, treatment with bypassing agents in the last 24 weeks; AND
5. For members **WITHOUT** inhibitors, member has severe hemophilia A (FVIII below 1%) or B (FIX equal to or below 2%); AND
6. Member's weight and bleeding history are documented; AND
7. Member will discontinue any other prophylactic therapy being used for hemophilia.
8. **Dosage allowed/Quantity limit:** Administer subcutaneously once daily as follows:

Day 1: Loading dose of 1 mg/kg
Day 2: 0.2 mg/kg once daily until individualization of maintenance dose
4 weeks after initiation: Measure Alhemo plasma concentration
No later than 8 weeks after initiation: Individualize maintenance dose based on Alhemo plasma concentration:

 - <200 ng/mL: 0.25 mg/kg
 - 200 to 4,000 ng/mL: 0.2 mg/kg
 - >4,000 ng/mL: 0.15 mg/kg

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

For **reauthorization**:

1. Chart notes must document a positive clinical response to treatment, such as fewer bleeding episodes compared to no prophylactic treatment; AND
2. Alhemo plasma concentration is being maintained above 200 ng/mL.

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

Qfitlia (fitusiran)

For **initial** authorization:

1. Member is at least 12 years of age; AND
2. Medication is being prescribed by or in consultation with a hematologist; AND
3. Member has diagnosis of severe congenital hemophilia A or B; AND
4. Member has documentation of antithrombin (AT) activity > 60% prior to treatment initiation; AND
5. Provider attests member will discontinue prophylactic therapy being used for hemophilia; AND
6. Chart notes include documentation of bleeding history.
7. **Dosage allowed/Quantity limit:** initial dose of 50 mg subcutaneously once every 2 months. Maintain AT activity between 15-35% by adjusting the dose and/or frequency of administration per package insert. Quantity limit: 1 pen or vial per 28 days.

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

For **reauthorization**:

1. Chart notes must show member experienced a reduction in bleeding episodes compared to baseline; AND
2. Documentation that member's target AT activity is between 15 and 35%

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, Ceprotrin

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	<ul style="list-style-type: none"> • On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes
	Afstyla	<ul style="list-style-type: none"> • On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes
	Kovaltry	<ul style="list-style-type: none"> • On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes
	Novoeight	<ul style="list-style-type: none"> • On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes
	Nuwiq Recombinate	<ul style="list-style-type: none"> • On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes

	Xyntha	<ul style="list-style-type: none"> 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	<ul style="list-style-type: none"> On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Altuviiiio	<ul style="list-style-type: none"> On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Eloctate	<ul style="list-style-type: none"> On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Esperoct	<ul style="list-style-type: none"> On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Jivi	<ul style="list-style-type: none"> On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
Non-Factor (Hemophilia A)	Hemlibra	<ul style="list-style-type: none"> 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
Recombinant Factor IX (Hemophilia B)	Benefix	Hemophilia B (congenital factor IX deficiency) for: <ul style="list-style-type: none"> On-demand treatment and control of bleeding episodes Perioperative management of bleeding Routine prophylaxis to reduce the frequency of bleeding episodes
	Ixinity	<ul style="list-style-type: none"> On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Rixubis	<ul style="list-style-type: none"> 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)	Alprolix	<ul style="list-style-type: none"> On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Idelvion	<ul style="list-style-type: none"> On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	Rebinyon	<ul style="list-style-type: none"> On-demand treatment and control of bleeding episodes Perioperative management Prevention and control of bleeding episodes
Plasma-Derived Factor IX (Hemophilia B)	AlphaNine SD	<ul style="list-style-type: none"> Prevention and control of bleeding episodes
Factor IX Complex (Hemophilia B)	Profilnine SD	<ul style="list-style-type: none"> Prevention and control of bleeding episodes
von Willebrand Factor/Coagulation Factor VIII Complex (Human)	Alphanate	<ul style="list-style-type: none"> Control and prevention of bleeding in patients with hemophilia A 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
	Humate-P	Hemophilia A <ul style="list-style-type: none"> Treatment and prevention of bleeding in adults Von Willebrand disease <ul style="list-style-type: none"> Treatment of spontaneous and trauma-induced bleeding episodes Perioperative management

	Wilate	<p>Children and adults with von Willebrand disease for:</p> <ul style="list-style-type: none"> On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes <p>Adolescents and adults with hemophilia A for:</p> <ul style="list-style-type: none"> On-demand treatment and control of bleeding episodes Routine prophylaxis to reduce the frequency of bleeding episodes
vonWillebrand Recombinant Factor	Vonvendi	<p>Adults with von Willebrand disease for:</p> <ul style="list-style-type: none"> On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes in patients with severe Type 3 von Willebrand disease receiving on-demand therapy.
Bypassing Agent	Feiba	<p>Hemophilia A and B with inhibitors for:</p> <ul style="list-style-type: none"> On-demand treatment and control of bleeding episodes Perioperative management Routine prophylaxis to reduce the frequency of bleeding episodes
	NovoSeven RT	<ul style="list-style-type: none"> Treatment of bleeding episodes and peri-operative management in 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	<ul style="list-style-type: none"> On-demand treatment of bleeding episodes in adults and adolescents with hemophilia A or B with inhibitors
Miscellaneous Factor	Obizur	<ul style="list-style-type: none"> On-demand treatment of bleeding episodes in adults with acquired hemophilia A
	Coagadex	<p>Hereditary Factor X deficiency for:</p> <ul style="list-style-type: none"> 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	<ul style="list-style-type: none"> Routine prophylactic treatment and peri-operative management of surgical bleeding in patients with congenital Factor XIII deficiency
	Tretten	<ul style="list-style-type: none"> Prophylaxis of bleeding in patients with congenital Factor XIII A-Subunit deficiency
	Fibryga	<ul style="list-style-type: none"> Treatment of acute bleeding episodes in adults and children with congenital fibrinogen deficiency, including afibrinogenemia and hypofibrinogenemia Fibrinogen supplementation in bleeding patients with acquired fibrinogen deficiency
	RiaSTAP	<ul style="list-style-type: none"> Treatment of acute bleeding episodes in adults and children with congenital fibrinogen deficiency, including afibrinogenemia and hypofibrinogenemia
Antithrombin	ATryn	<ul style="list-style-type: none"> <u>Prevention</u> of peri-operative and peri-partum thromboembolic events in patients with hereditary antithrombin deficiency
Protein C Concentrate	Ceprotrin	<ul style="list-style-type: none"> 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 Altuviio 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 Altuviio.
10/15/2024	Added acquired fibrinogen deficiency indication for Fibryga. Added criteria for Hympavzi.
01/03/2025	Updated references. Added criteria section for Alhemo.
04/14/2025	Added Qfitlia criteria section. Updated Hympavzi, Hemlibra and Alhemo to pharmacy or medical benefit; removed Hemofil and Recombinate from policy per discontinuation.
06/19/2025	Updated references; reduced age limit from 12 to 7 years of age per PI for Jivi

08/15/2025

Added criteria for patients who are not on inhibitors per label update, specified bypassing agent criteria is only for patients on inhibitors

References:

1. Advate [package insert]. Westlake Village, CA: Baxalta US Inc; 2018.
2. Adynovate [package insert]. Westlake Village, CA: Baxalta US Inc; 2021.
3. Afstylia [package insert]. Kankakee, IL: CSL Behring LLC; 2021.
4. Alhemo [prescribing information]. Novo Nordisk Inc.; 2025.
5. Alphanate [package insert]. Los Angeles, CA: Grifols Biologicals Inc.; 2021.
6. Alphanine SD [package insert]. Los Angeles, CA: Grifols Biologicals Inc.; 2017.
7. Alprolix [package insert]. Cambridge, MA: Biogen Inc.; 2020.
8. Altuviiiio [package insert]. Waltham, MA: Bioverativ Therapeutics Inc.; 2024.
9. ATryn [package insert]. Framingham, MA: rEVO Biologics, Inc.; 2013.
10. Benefix [package insert]. Philadelphia, PA: Wyeth Pharmaceuticals Inc.; 2021.
11. Ceprotin [package insert]. Lexington, MA: Baxalta US Inc.; 2021.
12. Coagadex [package insert]. Durham, NC: Bio Products Laboratory USA, Inc.; 2023.
13. Corifact [package insert]. Kankakee, IL: CSL Behring LLC; 2019.
14. Eloctate [package insert]. Waltham, MA: Bioverativ Therapeutics Inc.; 2020.
15. Esperoct [package insert]. Plainsboro, NJ: Novo Nordisk Inc.; 2022.
16. Feiba [package insert]. Westlake Village, CA: Baxter Healthcare Corporation; 2020.
17. Fibryga [package insert]. Paramus, NJ: Octapharma USA, Inc.; 2024.
18. Hemlibra [package insert]. South San Francisco, CA: Genentech, Inc.; 2022.
19. Hemofil M [package insert]. Lexington, MA: Baxalta US Inc.; 2018.
20. Humate-P [package insert]. Kankakee, IL: CSL Behring LLC; 2020.
21. Hymoviz [package insert]. New York, NY: Pfizer US Inc; 2024.
22. Idelvion [package insert]. Kankakee, IL: CSL Behring LLC; 2021.
23. Ixinity [package insert]. Berwyn, PA: Aptevo BioTherapeutics LLC; 2024.
24. Jivi [package insert]. Whippany, NJ: Bayer HealthCare LLC; 2025.
25. Koate-DVI [package insert]. Los Angeles, CA: Grifols Biologicals Inc.; 2012.
26. Kovaltry [package insert]. Whippany, NJ: Bayer HealthCare LLC; 2021.
27. Novoeight [package insert]. Plainsboro, NJ: Novo Nordisk Inc.; 2018.
28. Novoseven RT [package insert]. Bagsvaerd, Denmark: Novo Nordisk A/S; 2020.
29. NuwiQ [package insert]. Hoboken, NJ: Octapharma USA Inc.; 2021.
30. Obizur [package insert]. Westlake Village, CA: Baxter Healthcare Corporation; 2021.
31. Profilnine [package insert]. Los Angeles, CA: Grifols Biologicals Inc.; 2010.
32. Rebinyn [package insert]. Plainsboro, NJ: Novo Nordisk Inc.; 2022.
33. Recombinate [package insert]. Westlake Village, CA: Baxter Healthcare Corporation; 2018.
34. RiaSTAP [package insert]. Kankakee, IL: CSL Behring LLC; 2021.
35. Rixubis [package insert]. Westlake Village, CA: Baxalta US Inc.; 2020.
36. Qfitlia [package insert]. Genzyme Corporation; 2025.
37. Sevenfact [package insert]. Les Ulis, Franc: HEMA Biologics; 2020.
38. Tretten [package insert]. Bagsvaerd, Denmark: Novo Nordisk A/S; 2020.
39. VonVendi [package insert]. Westlake Village, CA: Baxalta US Inc.; 2022.
40. Wilate [package insert]. Hoboken, NJ: Octapharma USA Inc.; 2023.
41. Xyntha [package insert]. Philadelphia, PA: Wyeth Pharmaceuticals Inc.; 2022.
42. Srivastava A, Santagostino E, Dougall A, et al. WFH Guidelines for the Management of Hemophilia, 3rd edition. *Haemophilia*. 2020;26 Suppl 6:1-158. doi:10.1111/hae.14046
43. Oldenburg J. Optimal treatment strategies for hemophilia: achievements and limitations of current prophylactic regimens. *Blood*. 2015;125(13):2038-2044.
44. Hay CR. Immune tolerance induction: current status. *Hematology Education*. 2013;7:87-92
45. DiMichele DM, Hoots WK, Pipe SW, Rivard GE, Santagostino E. International workshop on immune tolerance induction: consensus recommendations. *Haemophilia*. 2007;13 Suppl 1:1-22.
46. Brackmann HH, White GC 2nd, Berntorp E, Andersen T, Escuriola-Ettingshausen C. Immune tolerance induction: What have we learned over time?. *Haemophilia*. 2018;24 Suppl 3:3-14.

47. Valentino LA, Kempton CL, Kruse-Jarres R, et al. US Guidelines for immune tolerance induction in patients with haemophilia a and inhibitors. *Haemophilia*. 2015;21(5):559-567.
48. Mahlangu J, Oldenburg J, Paz-Patel I, et al. Emicizumab Prophylaxis in Patients Who Have Hemophilia A without Inhibitors. *N Engl J Med*. 2018 Aug 30;379(9):811-822. doi: 10.1056/NEJMoa1803550.
49. Ng HJ, Lee LH. Recombinant activated clotting factor VII (rFVIIa) in the treatment of surgical and spontaneous bleeding episodes in hemophilic patients. *Vasc Health Risk Manag*. 2006;2(4):433-440. doi:10.2147/vhrm.2006.2.4.433.
50. Pipe S. Emicizumab subcutaneous dosing every 4 weeks is safe and efficacious in the control of bleeding in persons with haemophilia A with and without inhibitors – Results from the phase 3 HAVEN 4 study. Presented at the World Federation of Hemophilia World Congress in Glasgow, Scotland; May 20–24, 2018. WFH Oral Presentation.
51. Thornburg CD. How I approach: Previously untreated patients with severe congenital hemophilia A. *Pediatric blood & cancer*. 2018;65(12):e27466. doi:10.1002/pbc.27466.
52. Yada K, Nogami K. Spotlight on emicizumab in the management of hemophilia A: patient selection and special considerations. *J Blood Med*. 2019;10:171-181. Published 2019 Jul 2. doi:10.2147/JBM.S175952.
53. National Hemophilia Foundation (NHF). Recommendation on the Use and Management of Emicizumab-kxwh (Hemlibra) for Hemophilia A with and without Inhibitors. December 6, 2018.
54. Connell NT, Flood VH, Brignardello-Petersen R, et al. ASH ISTH NHF WFH 2021 guidelines on the management of von Willebrand disease. *Blood Adv*. 2021;5(1):301-325. doi:10.1182/bloodadvances.2020003264
55. Medical and Scientific Advisory Committee. MASAC recommendation regarding doses of clotting factor concentrate in the home. MASAC Document #242. June 2016.
56. Study of the Efficacy and Safety PF-06741086 in Adult and Teenage Participants with Severe Hemophilia A or Moderately Severe to Severe Hemophilia B. Clinicaltrials.gov identifier: NCT03938792. Updated October 10, 2024. Accessed October 18, 2024.
57. Matsushita T, Shapiro A, Abraham A, et al. Phase 3 Trial of Concizumab in Hemophilia with Inhibitors. *N Engl J Med*. 2023;389(9):783-794. doi:10.1056/NEJMoa2216455
58. MASAC Document 290 - MASAC Recommendations Concerning Products Licensed for the Treatment of Hemophilia and Selected Disorders of the Coagulation System. Available at: <https://www.bleeding.org/sites/default/files/document/files/MASAC-Products-Licensed.pdf>. Updated October 2, 2024. Accessed January 3, 2025.
59. Rezende SM, Neumann I, Angchaisuksiri P, et al. International Society on Thrombosis and Haemostasis clinical practice guideline for treatment of congenital hemophilia A and B based on the Grading of Recommendations Assessment, Development, and Evaluation methodology. *J Thromb Haemost*. 2024;22(9):2629-2652. doi:10.1016/j.jtha.2024.05.026
60. 2021 Georgia Code Title 33 – Insurance Chapter 20A - Managed Health Care Plans Article 2 - Patient's Right to Independent Review § 33-20A-31 Definitions. Justia US Law. Accessed April 25, 2023. <https://law.justia.com/codes/georgia/2021/title-33/chapter-20a/article-2/section-33-20a-31/>.

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