CareSource

PHARMACY POLICY STATEMENT
North Carolina Marketplace

DRUG NAME	Bleeding Disorder Agents
BILLING CODE	Must use valid J code – see Table A
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.

Bleeding Disorder Agents 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 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. Member's recent weight (kg), history of bleeds, and inhibitor status have been provided for review.
7. **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 titre 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 **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; 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 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, 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 **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 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 meet **one** of the following:
    a. Poor and/or frequent venous access AND risk outweighs benefit for obtaining a port or an alternative route of administration;
    b. Clinical documentation that prior prophylaxis with factor VIII (e.g., Advate, Adynovate, Eloctate, etc.) was ineffective for the prevention of bleeding episodes;
    c. Prescriber attested that member is not a candidate for factor VIII and the clinical rationale is strongly supported by chart notes; 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.

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	J Code
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	J7192
	Afstyla	• On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes	J7210
	Kovaltry	• On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes	J7211
	Novoeight	• On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes	J7182
	Nuwiq	• On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes	J7209
	Recombinate	• On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes	J7192
	Xyntha	• On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes	J7185
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	J7207
	Eloctate	• On-demand treatment and control of bleeding episodes • Perioperative management	J7205

		• 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	J7204
	Jivi	• On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes	J7208
Plasma-Derived Factor VIII (Hemophilia A)	Hemofil M	• Prevention and control of hemorrhagic episodes	J7190
	Koate	• Prevention and control of bleeding episodes	J7190
Non-Factor (Hemophilia A)	Hemlibra	• Routine prophylaxis to prevent or reduce the frequency of bleeding episodes in adults and pediatric patietns with **hemophilia A with or without factor VIII inhibitors**	J7170
Recombinant Factor IX (Hemophilia B)	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	J7195
	Ixinity	Adults and children ≥ 12 years of age with **hemophilia B** for: • On-demand treatment and control of bleeding episodes • Perioperative management Adults with **hemophilia B** for: • Routine prophylaxis to reduce the frequency of bleeding episodes	J7195
	Rixubis	• On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes	J7200
Extended Half-Life Recombinant Factor IX (Hemophilia B)	Alprolix	• On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes	J7201
	Idelvion	• On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes	J7202
	Rebinyn	• On-demand treatment and control of bleeding episodes • Perioperative management • Prevention and control of bleeding episodes	J7203

Plasma-Derived Factor IX (Hemophilia B)	AlphaNine SD	• Prevention and control of bleeding episodes	J7193
Factor IX Complex (Hemophilia B)	Profilnine SD	• Prevention and control of bleeding episodes	J7194
von Willebrand Factor/Coagulation Factor VIII Complex (Human)	Alphanate	• 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	J7186
	Humate-P	**Hemophilia A** • Treatment and prevention of bleeding in adults **Von Willebrand disease** • Treatment of spontaneous and trauma-induced bleeding episodes • Perioperative management	J7187
	Wilate	Children and adults with **von Willebrand disease** for: • On-demand treatment and control of bleeding episodes • Perioperative management Adolescents and adults with **hemophilia A** for: • On-demand treatment and control of bleeding episodes • Routine prophylaxis to reduce the frequency of bleeding episodes	J7183
vonWillebrand Recombinant Factor	Vonvendi	Adults with **von Willebrand disease** for: • 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.	J7179
Bypassing Agent	Feiba	**Hemophilia A and B with inhibitors** for: • On-demand treatment and control of bleeding episodes • Perioperative management • Routine prophylaxis to reduce the frequency of bleeding episodes	J7198
	NovoSeven RT	• 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	J7189

		• 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**	J7212
Miscellaneous Factor	Obizur	• On-demand treatment of bleeding episodes in adults with **acquired hemophilia A**	J7188
	Coagadex	**Hereditary Factor X deficiency** for: • 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 and moderate hereditary Factor X deficiency	J7175
	Corifact	• Routine prophylactic treatment and peri-operative management of surgical bleeding in patients with **congenital Factor XIII deficiency**	J7180
	Tretten	• Prophylaxis of bleeding in patients with **congenital Factor XIII A-Subunit deficiency**	J7181
	Fibryga	• Treatment of acute bleeding episodes in adults and children with **congenital fibrinogen deficiency**, including afibrinogenemia and hypofibrinogenemia	J7177
	RiaSTAP	• Treatment of acute bleeding episodes in adults and children with **congenital fibrinogen deficiency**, including afibrinogenemia and hypofibrinogenemia	J7178
Antithrombin	ATryn	• Prevention of peri-operative and peri-partum thromboembolic events in patients with **hereditary antithrombin deficiency**	J7196
Protein C Concentrate	Ceprotin	• Treatment and prevention of venous thrombosis and purpura fulminans in patients with **severe congenital Protein C deficiency**	J2724

CareSource considers Bleeding Disorder Agents 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.

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