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Department: PHARMACY	**MMC Review/ Approval Date(s):** 03/15/22	**Page(s): 17**

Policy Number: 040.003
Title: Coverage Determination Policy for Antihemophilic Agents

Regions: ☒ **Texas** ☐ **Florida** ☐ **Indiana** ☐ **New Jersey** ☒ **New Mexico**

Impacted Areas:

☒ Network Management/Provider Services	☒ Utilization Management
☐ Member services	☐ Case management
☐ Quality Management	☐ Disease management
☐ Credentialing	☒ Claims
☐ IT	☐ Human resources
☐ Administration	☐ Finance
☐ Compliance/delegation	☒ Pharmacy
	☐ ALL

Approved by: Vincent Huerta, MD, CHCQM Senior Medical Director	Vincent F Huerta Jr, MD, CHCQM Digitally signed by Vincent F Huerta Jr, MD, CHCQM Date: 2022.03.18 14:46:12 -05'00' *Signature of Approver*	03/18/2022 **Date:**
Reviewed by: Pathik Tripathi, Pharm.D, MS Director, Clinical Pharmacy	Pathik Tripathi Digitally signed by Pathik Tripathi Date: 2022.03.18 10:33:37 -05'00' *Signature of Reviewer*	03/18/2022 **Date:**

Available LCD/NCD/LCA:

- Local Coverage Determination (LCD): Hemophilia Factor Products (L35111)
- National Coverage Determination for Anti-Inhibitor Coagulant Complex (AICC) (110.3)
- LCA for Billing and Coding: HEMOPHIlia Factor Products : A56433

Disclaimer:
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Doctors helping patients for more than 25 years

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Coverage Policy Number: 040.003

Line of Business: Medicare Part B

Policy Type: Prior Authorization

Coverage Determination:

Initial/New Requests

Self-administered blood-clotting factors for hemophilia patients and items related to the administration of such factors are covered under Part B when ALL the following criteria exist:

1. The patient is diagnosed with any of the following:
    a. Factor VIII deficiency (classic hemophilia, hemophilia A).
    b. Factor IX deficiency (hemophilia B, Christmas disease, plasma thromboplastin component).
    c. Congenital factor XI deficiency (Hemophilia C).
    d. Von Willebrand's disease.
    e. Acquired hemophilia (acquired Factor VIII autoantibodies most frequently) and other coagulation factor deficiencies, intrinsic circulating anticoagulants, antibodies or inhibitors.
    f. Congenital deficiencies of other clotting factors (such as congenital afibrinogenemia and others).
2. The Factor is used to control bleeding associated with hemophilia.
3. The patient is competent to use such factors without medical or other supervision.
4. A profile of the patient's use and a prescription for supplies should be submitted with a beneficiary new to the Contractor or a newly enrolled beneficiary.

Feiba Anti-Inhibitor Coagulation Complex (AICC) (J7198)

AICC has been shown to be safe and effective and is covered when **ALL** of the following criteria are met:

1. Patient has diagnosis of hemophilia A or B
2. Patient has inhibitors to Factor VIII and IX
3. Has had major bleeding episodes
4. Has failed to respond to other less-expensive therapies

NovoSeven (J7189)

Factor VIIa (anti-hemophilic factor, recombinant) is covered when ALL of the following criteria are met:

1. Patient has one or more of the following indications:
    a. Treatment of bleeding episodes or perioperative management in hemophilia A or B with inhibitors
    b. Congenital Factor VII deficiency
    c. Glanzmann's thrombasthenia with refractoriness to platelet transfusions, with or without antibodies to platelets
    d. Treatment in bleeding episodes and perioperative management in adults with acquired hemophilia.
2. Initially administered under the supervision of a physician experienced in the treatment of bleeding disorders

3. Effectiveness monitored by hemostasis evaluations to provide a basis for modification of the treatment schedule.

PLEASE NOTE: NovoSeven is **NOT** covered for prophylaxis treatment other than for perioperative invasive procedures or surgery.

Hemlibra (J7170)

Emicizumab-kxwh [Hemlibra] is covered when the following criteria are met:

1. Diagnosis of severe hemophilia A
2. Documentation of endogenous factor VIII level less than 1% of normal factor VIII (< 0.01 IU/mL);

OR

1. Diagnosis of hemophilia A regardless of severity
2. Submission of medical records (e.g., chart notes, laboratory values) documenting a failure to meet clinical goals (e.g., continuation of spontaneous bleeds, inability to achieve appropriate trough level, previous history of inhibitors) after a trial of prophylactic factor VIII replacement products

OR

1. Diagnosis of hemophilia A
2. Patient has developed high-titer factor VIII inhibitors (≥ 5 Bethesda units [BU])
3. Prescribed for the prevention of bleeding episodes (i.e., routine prophylaxis)

Renewal/Continuation of Therapy Requests

RENEWAL requests for continued use of the above products will be approved if ALL of the following are met:

1. The patient has documentation of positive clinical response
2. The requested dosing regimen remains within the recommended dosing parameters
3. The patient still meets all of the indication-specific criteria above

FDA Approved Dose and Indication:

DRUG	INDICATION/DOSE
Feiba Anti-Inhibitor Coagulation Complex (AICC) (J7198)	**Hereditary factors IX & VIII deficiency disease with inhibitors** **Hemorrhage prophylaxis**- 85 units/kg IV every other day **Hemorrhage**: • Joint: 50 to 100 units/kg IV every 12 hours until reduction in pain and in acute disability • Mucous: 50 to 100 units/kg IV every 6 hours for at least 1 day or until bleeding resolves • Soft tissue: 100 units/kg IV every 12 hours until bleeding resolves • Severe: 100 units/kg IV every 6 to 12 hours until bleeding resolves **Surgical procedure**: • Preoperative dose: 50 to 100 units/kg IV immediately before surgery • Postoperative dose: 50 to 100 units/kg IV every 6 to 12 hours until bleeding resolves and healing is achieved
NovoSeven (J7189)	**Acquired hemophilia**: 70 to 90 mcg/kg slow IV bolus every 2 to 3 hours until hemostasis **Factor VII deficiency**: 15 to 30 mcg/kg IV bolus, then every 4 to 6 hours until hemostasis achieved **Glanzmann's thrombasthenia**: 90 mcg/kg IV bolus every 2 to 6 hours. Higher doses of 100 to 140 mcg/kg can be used **Hemophilia A or B**: • Hemorrhage: 90 mcg/kg IV bolus; repeat every 2 hours until hemostasis. At home treatment- 90 mcg/kg IV bolus followed by 15 to 16 mcg/kg/hr for 12 hours with a target trough blood factor VII level of 10 units/mL, OR 160 to 180 mcg/kg bolus followed by 30 mcg/kg/hr for 6 hours with a target trough blood factor VII level of 20 units/mL (off-label dosage) • Prophylaxis: Initial: 90 mcg/kg IV bolus immediately prior to intervention and repeat every 2 hours for duration of surgery. Minor post-surgery 90 mcg/kg IV bolus every 2 hours for first 48 hours, then every 2 to 6 hours until healing has occurred. Major post-surgery 90 mcg/kg IV bolus every 2 hours for 5 days then every 4 hours or by continuous infusion at 50 mcg/kg/hr until healing has occurred; additional bolus doses can be given
Hemlibra (J7170)	Hemophilia A Hemorrhage prophylaxis: Initial 3 mg/kg subQ once weekly for 4 weeks. Maintenance 1.5 mg/kg subQ once weekly OR 3 mg/kg every 2 weeks OR 6 mg/kg every 4 weeks; base dose selection on provider preference with consideration for regimens that may increase patient adherence.
Novoeight (J7182)	Hemophilia A Routine prophylaxis: 20-50 units/kg 3 times a week OR 20-40 units/kg every other day
Wilate (J7183)	Hemophilia A - Hemorrhage: • Minor: 30-40 units/kg every 12-24 hours for at least 1 day * • Moderate: 30-40 units/kg every 12-24 hours for at least 3-4 days * • Major: 35-50 units/kg every 12-24 hours for at least 3 to 4 days * • Life threatening: 35-50 units/kg every 8-24 hours until threat has resolved; *

	• Prophylaxis: 20-40 units/kg every 2-3 days * titrate dose and frequency based on clinical response and clinical condition, severity of deficiency, severity of hemorrhage, desired factor VIII level, and presence of inhibitor
Xyntha (J7185)	Hemophilia A Hemorrhage: • Minor: 10-20 units/kg every 12-24 hours • Moderate: 15-30 units/kg every 12-24 hours • Major: 30-50 units/kg every 8-24 hours Prophylaxis: • 30 international units/kg IV 3 times weekly Surgical prophylaxis: • Minor: 15-30 units/kg every 12-24 hours • Major: 30-50 units/kg every 8-24 hours
Humate P (J7186)	Hemophilia A Hemorrhage: • Minor: 15 units/kg for one dose then if needed 7.5 units/kg once or twice daily for 1 to 2 days • Moderate: 25 units/kg loading dose, then 15 units/kg every 8-12 hours for 1 to 2 days then once or twice daily • Life-threatening: 40-50 units/kg loading dose, then 20-25 units/kg every 8 hours for 7 days then once or twice daily for 7 days Von Willebrand disorder: See Micromedex for dosing
Rixubis (J7200)	**Hemophilia B** **Antihemophilic Agent**: Number of factor IX international units required = body weight (kg) multiplied by desired factor IX increase (% or international units/dL) multiplied by 1.1 dL/kg **Hemorrhage prophylaxis**: Previously treated patients: 40 to 60 international units/kg IV twice a week; adjust as necessary for individual age, bleeding pattern, and physical activity. **Perioperative care:** • Minor surgery: 30 to 60 international units/dL or 30% to 60% of normal factor IX level required IV every 24 hours for 1 or more days until healing is achieved • Major surgery: 80 to 100 international units/dL or 80 to 100% of normal factor IX level required IV every 8 to 24 hours for 7 to 10 days until bleeding stops and healing is achieved
Alprolix (J7201)	**Hemophilia B** **Hemorrhage:** • **Minor to moderate (30% to 60% of normal Factor IX level required)**: IV bolus infusion; repeat every 48 hours with further evidence of bleeding • **Major (80% to 100% of normal Factor IX level required)**: IV bolus infusion; consider repeat dose after 6 to 10 hours then every 24 hours for the first 3 days; after day 3 may decrease dose and increase interval to every 48 hours or longer until bleeding ceases and healing occurs Surgical prophylaxis and treatment: • Minor (50% to 80% of normal factor IX level required): Single IV bolus infusion, repeat as needed after 24 to 48 hours until bleeding ceases and healing occurs

	• Major (60% to 100% of normal factor IX initial level required): IV bolus infusion; consider repeat dose after 6 to 10 hours then every 24 hours for first 3 days; after day 3 may decrease dose and increase interval to every 48 hours or longer until bleeding ceases and healing occurs Prophylaxis of bleeding episodes: 50 international units/kg IV bolus infusion once weekly OR 100 international units/kg once IV bolus infusion every 10 days
Esperoct (J7204)	**Hemophilia A** **Hemorrhage:** • Minor-Moderate: 40 units/kg; for moderate, may repeat an additional dose after 24 hours • Major: 50 units/kg; may repeat doses every 24 hours **Perioperative:** 50 units/kg Prophylaxis : 50 units/kg every 4 days
Idelvion (J7202)	**Hemophilia B** **Hemorrhage prophylaxis:** initial 25 to 40 international units/kg IV infusion every 7 days; may switch to 50 to 75 international units/kg IV every 14 days **Hemorrhage**: • Minor to moderate: (30% to 60% of circulating Factor IX level required) IV infusion for at least 1 day, until bleeding stops and healing is achieved. May be repeated every 48 to 72 hours, a single dose should be sufficient for most bleeds • Major: (60% to 100% of circulating Factor IX level required) IV infusion every 48 to 72 hours for 7 to 14 days and until bleeding stops and healing is achieved Perioperative: • Minor: (50% to 80% of circulating factor IX level required) IV infusion for at least 1 day or until healing is achieved. May repeat every 48 to 72 hours • Major: (60% to 100% of circulating factor IX initial level required) IV infusion every 48 to 72 hours for the first week or until healing is achieved; continue 7 to 14 days, or until healing complete. Administer maintenance dose 1 to 2 times/week
Nuwiq (J7209)	**Hemorrhage:** • Minor: Increase in plasma level of antihemophilic factor of 20% to 40% of normal, repeat IV dose every 12 to 24 hours for at least 1 day • Moderate to Major: Increase in plasma level of antihemophilic factor of 30% to 60% of normal, repeat IV dose every 12 to 24 hours for 3 to 4 days • Life-threatening: : Increase in plasma level of antihemophilic factor of 60% to 100% of normal, every 8 to 24 hours until hemostasis is achieved **Hemorrhage prophylaxis:** 30 to 40 international units/kg IV every other day **Surgical procedure:** • Minor: Increase in plasma level of antihemophilic factor of 30% to 60% of normal; repeat IV infusions every 24 hours for at least 1 day • Major: Increase in plasma level of antihemophilic factor of 80% to 100% of normal; repeat IV infusions every 8 to 24 hours
Factor IX Complex Human (J7194)	**Hemophilia B** **Hemorrhage:** • Mild to moderate: raise plasma factor IX level to 20% to 30% in a single administration

	• Severe: raise plasma factor IX level to 30% to 50% administered daily **Surgery**: raise plasma factor IX level to 30% to 50%
Adynovate- Factor VIII pegylated recombinant (J7207)	**Hemophilia A** **Perioperative:** • **Minor:** 30 to 50 international units/kg • **Major:** 40 to 60 international units/kg **Prophylaxis:** • **40 to 50 international units/kg twice a week** **Hemorrhage:** • **Minor: 10 to 20 international units/kg IV every 12 to 24 hours** • **Moderate: 15 to 30 international units/kg IV every 12 to 24 hours** • **Major: 30 to 50 international units/kg IV every 8 to 24 hours**
Eloctate (J7205)	**Hemophilia A** **Hemorrhage:** • Minor-moderate: 20-30 units/kg IV every 24 to 48 hours until bleeding resolved • Major: 40 to 50 international units/kg IV every 12 to 24 hours for 7 to 10 days or until bleeding resolved **Perioperative:** • Minor: 25 to 40 units/kg IV every 24 hours for at least 1 day • Major: 40 to 60 international units/kg IV preoperatively, followed by 40 to 50 international units/kg IV after 8 to 24 hours and then every 24 hours **Hemorrhage prophylaxis:** Initial 50 units/kg every 4 days. Maintenance 25-65 units/kg every 3-5 days
Thrombate III (J7197)	**Antithrombin III deficiency, Hereditary - Thromboembolic disorder** **Peripartum/perioperative Prophylaxis:** Loading dose, (120% - baseline % x body weight (kg))/1.4% units to target AT level 120% of normal. Maintenance dosage, loading dose x 0.6 units (target AT level 80% to 120% of normal) every 24 hours **Treatment and prophylaxis:** Loading dose, (120% - baseline % x body weight (kg))/1.4% units to target AT level 120% of normal. Maintenance dosage, loading dose x 0.6 units (target AT level 80% to 120% of normal) every 24 hours
Factor IX recombinant nos (J7195)	**BeneFIX- Hemophilia B** • Mild hemorrhage: 20% to 30% of normal factor IX level or 20 to 30 international units/dL factor IX activity required IV every 12 to 24 hours • Moderate hemorrhage: 25% to 50% of normal factor IX level or 25 to 50 international units/dL activity required IV every 12 to 24 hours for 2 to 7 days • Major hemorrhage: 50% to 100% of normal factor IX level or 50 to 100 international units/dL activity required IV every 12 to 24 hours for 7 to 10 days • Long-term prophylaxis: 100 international units/kg IV once weekly **Ixinity- Hemophilia B** • Minor hemorrhage: 30% to 60% of normal factor IX level or 30 to 60 international units/dL factor IX activity required IV every 24 hours • Moderate hemorrhage: 40% to 60% of normal factor IX level or 40 to 60 international units/dL factor IX activity required IV every 24 hours

	• Major or life-threatening hemorrhage: 60% to 100% of normal factor IX level or 60 to 100 international units/dL factor IX activity required IV every 12 to 24 hours • Hemorrhage prophylaxis: Previously treated patients: 40 to 70 international units/kg IV twice weekly • Minor Surgery Pre-op: 50 to 80 international units/dL or 50% to 80% of normal factor IX level required • Major surgery Pre-op: 60 to 80 international units/dL or 60% to 80% of normal factor IX level required
Profilnine (J7194)	Hemophilia B • Mild to moderate hemorrhage: raise plasma factor IX level to 20% to 30% in a single administration. • Severe hemorrhage: raise plasma factor IX level to 30% to 50% administered daily. • Surgery: raise plasma factor IX level to 30% to 50% administered for at least 1 week following surgery. • Dental extractions: raise plasma factor IX level to 50% administered just prior to procedure and give additional doses as needed
Factor IX non-recombinant (J7193)	**Hemophilia B** **Alphanine SD** • Mild hemorrhage: 20 to 30 international units/kg IV twice daily. • Moderate hemorrhage: 25 to 50 international units/kg IV twice a day. • Major hemorrhage: 30 to 50 international units/kilogram IV twice a day for at least 3 to 5 days. Followed by 20 international units/kg IV twice a day. • Surgery: 50 to 100 international units/kg IV twice daily for 7 to 10 days. **Mononine** • Mild hemorrhage: 20 to 30 international units/kg IV once and repeated in 24 hours if needed. • Major trauma or surgery: up to 75 international units/kg IV every 18 to 30 hours.
Factor VIII recombinant nos (J7192)	**Hemophilia A** **Kovaltry** • Minor hemorrhage: Increase in plasma level of antihemophilic factor of 20% to 40% of normal, repeat IV dose every 12 to 24 hours. • Moderate hemorrhage: Increase in plasma level of antihemophilic factor of 30% to 60% of normal, repeat IV dose every 12 to 24. • Major hemorrhage: Increase in plasma level of antihemophilic factor of 60% to 100% of normal, every 8 to 24 hours. • Hemorrhage prophylaxis: 20 to 40 international units/kg IV 2 or 3 times per week. • Minor surgery: Increase in plasma level of antihemophilic factor of 30% to 60% of normal; repeat IV infusions every 24 hours. • Major surgery: Increase in plasma level of antihemophilic factor of 80% to 100% of normal; repeat IV infusions every 8 to 24 hours. **Recombinate** • Minor hemorrhage, increase in plasma level of antihemophilic factor of 20% to 40% of normal: Begin IV infusions every 12 to 24 hours. • Moderate hemorrhage: Increase in plasma level of antihemophilic factor of 30% to 60% of normal, repeat IV infusion every 12 to 24 hours.

	• Major hemorrhage: Increase in plasma level of antihemophilic factor of 60% to 100% of normal, repeat IV infusions every 8 to 24 hours. • Minor surgery: Single IV bolus infusion to achieve increase in plasma level of antihemophilic factor of 60% to 80% of normal. • Major surgery: Increase in plasma level of antihemophilic factor of 80% to 100% of normal (pre-and-post operative); repeat IV infusions every 8 to 24 hours.
Koate Factor VIII human (J7190)	**Hemophilia A** **Hemorrhage:** • Mild: single dose of 10 units/kg • Moderate: 15-25 units/kg. Repeat with 10-15 units/kg every 8-12 hours if evidence of further bleeding. • Severe: initially 40 to 50 units/kg; maintenance dose 20 to 25 units/kg every 8 to 12 hours. **Surgery:** • Major procedures: preoperatively 50 units/kg. Repeat infusions every 6 to 12 hours initially as needed and for a total of 10 to 14 days.
AlphanateAntihemophilic VIII/VWF complex (J7186)	**Hemophilia A** **Hemorrhage:** • Minor: 15 units/kg twice daily (1 to 2 days) • Moderate: 25 units/kg twice daily (2 to 7 days) • Major: 40 to 50 units/kg twice daily for at least 3 to 5 days then 25 units/kg twice daily **Von Willebrand disorder - Surgery** • **Loading dose, 60 international units VWF:RCo/kg IV** • **Minor: maintenance dose, 40 to 60 international units VWF:RCo/kg IV every 8 to 12 hours** • **Major: maintenance dose, 40 to 60 international units VWF:RCo/kg IV every 8 to 12 hours**

General Background:

Hemophilia is a hereditary blood disease characterized by greatly prolonged coagulation time. The blood fails to clot and abnormal bleeding occurs. It is a sex-linked hereditary trait transmitted by normal heterozygous females who carry the recessive gene. It occurs almost exclusively in males. For purposes of Medicare coverage, hemophilia encompasses Factor VIII deficiency (classic hemophilia, hemophilia A), Factor IX deficiency (hemophilia B, Christmas disease, plasma thromboplastin component), and von Willebrand's disease. Approximately 80% of those with hemophilia have type A, and both are associated with recurrent, spontaneous, and traumatic hemarthrosis.

The frequency and severity of hemorrhagic events induced by hemophilia are related to the amount of coagulation factor in the blood. Those with mild hemophilia (defined as having from 5% to 40% of normal coagulation factor activity) experience complications only after having undergone surgery or experiencing a major physical trauma. Those with moderate hemophilia (from 1% to 5% of coagulation factor activity) experience some spontaneous hemorrhage but normally exhibit bleeding provoked by trauma. Those with severe hemophilia (less than 1% of coagulation factor activity) exhibit spontaneous hemarthrosis and bleeding. Treatment for these patients is dependent on the severity of the disease and may include the administration of blood clotting factors such as Factor VIII, Factor IX, Factor VIIa and, Anti-inhibitors to control the bleeding.

Medicare provides coverage of these factor products through Part A and B coverage. In Part B, Medicare provides coverage in two manners, one of an 'incident to' event where the provider has a cost of the factor and administers, whereby the claim will demonstrate the factor product code and administration codes. Medicare also provides coverage for self-administered blood-clotting factors for hemophilia patients who are competent to use such factors to control bleeding without medical supervision. Medicare covers blood clotting factors for the following conditions:

- Factor VIII deficiency (classic hemophilia, hemophilia A)
- Factor IX deficiency (hemophilia B, Christmas disease, plasma thromboplastin component)
- Congenital factor XI deficiency (Hemophilia C)
- Von Willebrand's disease
- Acquired hemophilia (acquired Factor VIII autoantibodies most frequently) and other coagulation factor deficiencies, intrinsic circulating anticoagulants, antibodies or inhibitors.
- Congenital deficiencies of other clotting factors (such as congenital afibrinogenemia and others).

Anti-Inhibitor Coagulation Complex (AICC) (Feiba, VH Immuno, Autoplex or Hemophilia clotting factor) is a drug used to treat hemophilia in patients with Factor VIII inhibitor antibodies. AICC has been shown to be safe and effective and is covered when furnished to patients with hemophilia A or B and inhibitor antibodies to Factor VIII who have major bleeding episodes and who fail to respond to other less-expensive therapies.

Factor VIIa (anti-hemophilic factor, recombinant) (NovoSeven) is indicated for the treatment of bleeding episodes or perioperative management in hemophilia A or B with inhibitors, congenital Factor VII deficiency and Glanzmann's thromboasthenia with refractoriness to platelet transfusions, with or without antibodies to platelets. NovoSeven is also labeled for treatment in bleeding episodes and perioperative management in adults with acquired hemophilia. NovoSeven is not labeled for prophylaxis treatment other than for perioperative invasive procedures or surgery. NovoSeven, as noted in the Prescribing Information for the product, should be administered to patients initially under the supervision of a physician experienced in the treatment of bleeding disorders. Effectiveness of NovoSeven should be monitored by hemostasis evaluations to provide a basis for modification of the treatment schedule.

Emicizumab-kxwh (Hemlibra®) is approved by the FDA as the originator biological product for routine prophylaxis to prevent or reduce the frequency of bleeding events in adult and pediatric patients (newborn and older) with congenital factor VIII deficiency (hemophilia A) with or without factor VIII inhibitors

Clinical Evidence:

Antihemophilic factor is usually indicated for hemophilia when a bleeding episode arises (demand treatment) or when bleeding is anticipated or likely (prophylactic treatment). Primary prophylactic therapy may be indicated for patients with severe hemophilia A or B who have less than 1 percent of normal factor (less than 0.01 IU/mL (National Hemophilia Foundation, 2001). Primary prophylactic therapy should be instituted early, prior to the onset of frequent bleeding, with the aim of keeping the trough factor or Factor VIII or Factor IX level above 1 percent between doses (National Hemophilia Foundation, 2001). In some cases, continuous prophylactic therapy may be indicated in persons with hemophilia A or hemophilia B that is not severe (i.e., hemophiliacs with more than 1 percent of normal factor levels) who have repeated episodes of spontaneous bleeding. Inhibitors are antibodies that neutralize Factor VIII and can render replacement therapy ineffective. They are found more commonly in patients with moderate to severe hemophilia (up to 30 percent of those with severe disease) who have received significant amounts of replacement therapy. Immune tolerance strategies in those with identified inhibitors also have been successful. Assuming no anamnestic response, low-titer inhibitors occasionally can be overcome with high doses of Factor VIII. Recombinant human coagulation Factor VIIa (rFVIIa) is indicated for the treatment of patients with bleeding episodes and for the prevention of bleeding in surgical interventions or invasive procedures in patients with hemophilia A or B with inhibitors to Factor VIII or Factor IX. High-titer inhibitors have been treated with variable success using porcine Factor VIII, Factor IX complex concentrates, recombinant Factor VIII, and exchange plasma pheresis. Anti-Inhibitor Coagulant Complex (AICC) is a drug used to treat hemophilia in patients with Factor VIII inhibitor antibodies. AICC has been shown to be safe and effective and is covered by Medicare when furnished to patients with hemophilia A and inhibitor antibodies to Factor VIII who have major bleeding episodes and who fail to respond to other less expensive therapies.

Immune tolerance induction is designed to overcome the effects of antihemophilic factor or Factor IX inhibitors in certain hemophiliac patients, thus restoring effectiveness of antihemophilic factor or Factor IX therapy to resolve active bleeding in these patients. It consists of administration of very high doses of anti-hemophilic factor or Factor IX over an extended period of time.

HCPCS Code:

Description	HCPCS Code
Emicizumab-Kxwh 0.5mg	J7170
Factor XIII anti-hem factor, 1 IU	J7180
Factor VIII recombinant Novoeight, per IU	J7182
Wilate injection, von Willebrand factor complex (human) 1 IU vWF:RCo	J7183
Xyntha injection, factor VIII (antihemophilic factor, recombinant) per IU	J7185
Antihemophilic VIII/VWF complex (human), per factor VIII IU	J7186
Humate-P	J7187
NovoSeven RT, Factor VIIa (antihemophilic factor, recombinant), 1 mcg	J7189
Factor VIII (antihemophilic factor, human), per IU	J7190
Factor VIII recombinant nos, per IU	J7192
Factor IX non-recombinant nos, per IU	J7193
Factor IX Complex, per IU	J7194
Factor IX Recombinant nos, per IU	J7195
Antithrombin III, per IU	J7197
Anti-Inhibitor, per IU	J7198
Factor IX (antihemophilic factor, Recombinant) Rixubis, per IU	J7200
Factor IX, FC fusion protein Recombinant, Alprolix per IU	J7201
Factor IX Idelvion	J7202
Recombinant Esperoct per iu	J7204
Factor VIII FC Fusion Recombinant	J7205
Factor VIII Pegylated Recombinant	J7207
Factor VIII Nuwiq Recombinant	J7209

Acronyms:

NCD = National Coverage Determination; LCD = Local Coverage Determination; CMS = Centers for Medicare and Medicaid Services; FDA = Food and Drug Administration; AICC = Anti-Inhibitor Coagulation Complex

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2. CMS IOM Publication 100-03, Medicare Claims Processing Manual. Chapter 17; § 40 Discarded Drugs and Biologicals, § 80.4-80.4.1 Billing for Hemophilia Clotting Factors/Clotting Factor Furnishing Fee, § 90.2 Drugs, Biologicals, and Radiopharmaceuticals. Accessed at https://www.cms.gov/Regulations-and-Guidance/Guidance/Manuals/Downloads/clm104c17.pdf. Accessed on 12/31/20.
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Policy History/Revision Information:

Date Revised	Type of Changes (Significant or Minor)	List Significant Changes and/or Status of policy
1/18/19	Significant	New coverage criteria adopted from NCD and LCD. - S. Younts, PharmD, MPH, BCPS
12/10/19	Significant	Added New Mexico and Indiana to the regions section and NCD/LCD/LCA information under attachments with hyperlinks. Added coverage for Hemophilia C. Added renewal section. Added information on Hemlibra to the background information section. Added coverage criteria for Hemlibra. Updated references. – Eric McDermott, PharmD
12/9/21	Significant	Updated to new formatting. Tabularized the code list. Included FDA approved doses and indications. Added references. Hyperlinks inserted for table on content. Nana Brobbey, PharmD
02/18/22	Minor	Policy title updated from Hemophilia factors to Antihemophilic agents, Added links for LCD/LCA, updated criteria information for Feiba from Micromedex, updated some dosing from Micromedex, updated references. - Cordelia Osidele, PharmD