EmblemHealth

Clotting Disorder Therapy

Effective Date: February 20, 2021	Number: MG.MM.PH.25

Medical Guideline Disclaimer

 The treating physician or primary care provider must submit to EmblemHealth the clinical evidence that the patient meets the criteria for the treatment or surgical procedure. Without this documentation and information, EmblemHealth will not be able to properly review the request for prior authorization. The clinical review criteria expressed below reflects how EmblemHealth determines whether certain services or supplies are medically necessary. EmblemHealth established the clinical review criteria based upon a review of currently available clinical information (including clinical outcome studies in the peer-reviewed published medical literature, regulatory status of the technology, evidence-based guidelines of public health and health research agencies, evidence-based guidelines and positions of leading national health professional organizations, views of physicians practicing in relevant clinical areas, and other relevant factors). EmblemHealth expressly reserves the right to revise these conclusions as clinical information changes, and welcomes further relevant information. Each benefit program defines which services are covered. The conclusion that a particular service or supply is medically necessary does not constitute a representation or warranty that this service or supply is covered and/or paid for by EmblemHealth, as some programs exclude coverage for services or supplies that EmblemHealth considers medically necessary. If there is a discrepancy between this guideline and a member's benefits program, the benefits program will govern. In addition, coverage may be mandated by applicable legal requirements of a state, the Federal Government or the Centers for Medicare & Medicaid Services (CMS) for Medicare and Medicaid members. All coding and web site links are accurate at time of publication. EmblemHealth Services Company LLC, ("EmblemHealth") has adopted the herein policy in providing management, administrative and other services to, EmblemHealth Plan, Inc., EmblemHealth Insurance Company, EmblemHealth Services Company, LLC and Health Insurance Plan of Greater New York (HIP), ConnectiCare, Inc., ConnectiCare Insurance Company, Inc. ConnectiCare Benefits, Inc., and ConnectiCare of Massachusetts, Inc. related to health benefit plans offered by these entities. All of the aforementioned entities are affiliated companies under common control of EmblemHealth Inc.

Dosage and Administration

Refer to drug-specific labeling for appropriate dosing and administration

Applicable Products

Criteria I	AlphaNine SD, Alprolix, Bebulin, BeneFIX, Idelvion, Profilnine SD, Mononine, Rixubis, IXINITY, and Rebinyn
Criteria II	Alphanate, Humate-P
Criteria III	Advate, Adynovate, Eloctate, Helixate FS, Hemofil M, Koate-DVI, Kogenate FS, Monoclate-P, Novoeight, Recombinate, Xyntha, NUWIQ, Afstyla, Kovaltry, Jivi, Esperoct
Criteria IV	Feiba NF
Criteria V	Novoseven RT
Criteria VI	Obizur
Criteria VII	Tretten
Criteria VIII	Corifact
Criteria IX	Wilate
Criteria X	Hemlibra

Criteria XI	Vonvendi
Criteria XII	Coagadex
Criteria XIII	Sevenfact

Definitions

Hemophilia A AKA factor VIII (FVIII) deficiency or classic hemophilia	Genetic disorder caused by missing or defective factor VIII, a clotting protein.
Hemophilia B AKA factor IX (FIX) deficiency or Christmas disease	Genetic disorder caused by missing or defective factor IX, a clotting protein.
Congenital factor VII deficiency AKA Hageman Factor	FXII deficiency is inherited in an autosomal recessive fashion, meaning both parents must carry the gene to pass it on to their children
Glanzmann Thrombasthenia	Genetic disorder in which the platelets have qualitative or quantitative deficiencies of the fibrinogen receptor αIIbβ3
Von Willebrand disease (VWD)	Genetic disorder caused by missing or defective von Willebrand factor (VWF), a clotting protein. VWF binds factor VIII, a key clotting protein, and platelets in blood vessel walls, which help form a platelet plug during the clotting process.

The medications listed in this policy are not covered by the pharmacy benefit. Human Antihemophilic Factor products are covered by the medical benefit and must be obtained by Accredo, our preferred medical specialty provider. Exceptions to Accredo fulfillment may be considered with urgent circumstances on an individual basis.

Specialty trained clinicians at Accredo's Therapeutic Resource Center for bleeding disorders will provide personalized care, and support services to minimize drug-related adverse episodes, and/ or gaps in care. Accredo will perform assay management, coordinate delivery based on site of care, provide financial assistance coordination, and emphasize patient empowerment strategies to improve adherence.

Accredo can be reached by phone at 866.712.5007 or fax 800.330.07566

Guideline

Hemophilia medications are considered medically necessary when the following product-specific criteria are met:

Criteria I — AlphaNine SD, Alprolix, Bebulin, BeneFIX, Idelvion, Profilnine SD, Mononine, Rixubis, IXINITY, Rebinyn

1. Hemophilia B
    a. Congenital factor IX deficiency confirmed by blood coagulation testing; AND

b. Used to treat at least 1:
    i. Control and prevention of acute bleeding episodes (hemorrhage)
    ii. Routine prophylaxis to prevent or reduce frequency of bleeding episodes (applicable to AlphaNine SD, Alprolix, Bebulin, BeneFIX, Profilnine SD, Mononine, Rixubis, and IXINITY only)
    iii. Perioperative management; AND
c. Not used for induction of immune tolerance in members with Hemophilia (applicable to Alprolix, Rixubis, Idelvion, IXINITY and Rebinyn only)

Criteria II — Alphanate, Humate-P

1. Hemophilia A
    a. Congenital factor VIII deficiency confirmed by blood coagulation testing; AND
    b. Used to treat at least 1:
        i. Control and prevention of acute bleeding episodes (hemorrhage)
        ii. Routine prophylaxis to prevent or reduce the frequency of bleeding episodes
        iii. Perioperative management
2. Von Willebrand disease (VWD)
    a. Von Willebrand disease confirmed by blood coagulation and von Willebrand factor testing; AND
    b. Used as surgical bleeding prophylaxis in members with VWD in whom desmopressin is either ineffective or contraindicated; OR to control episodes of acute bleeding (Humate-P only); AND
    c. Alphanate is not indicated for members with severe (type 3) VWD undergoing major surgery

Criteria III — Advate, Adynovate, Eloctate, Helixate FS, Hemofil M, Koate-DVI, Kogenate FS, Monoclate-P, Novoeight, Recombinate, Xyntha, NUWIQ, Afstyla, Kovaltry, Jivi, Esperoct

1. Hemophilia A
    a. Congenital factor VIII deficiency confirmed by blood coagulation testing; AND
    b. Used to treat at least 1:
        i. Control and prevention of acute bleeding episodes (hemorrhage)
        ii. Routine prophylaxis to prevent or reduce frequency of episodic bleeding
        iii. Perioperative management

Criteria IV — Feiba NF

1. Hemophilia A

a. Congenital factor VIII deficiency confirmed by blood coagulation testing; AND
b. Confirmation that member has Factor VIII inhibitors; AND
d. Used to treat at least 1:
    i. Control and prevention of acute episodic bleeding (hemorrhage)
    ii. Routine prophylaxis to prevent or reduce the frequency of episodic bleeding

2. Hemophilia B
    a. Congenital factor IX deficiency confirmed by blood coagulation testing; AND
    b. Confirmation that member has Factor IX inhibitors; AND
    e. Used to treat at least 1:
        i. Control and prevention of acute episodic bleeding (hemorrhage)
        ii. Routine prophylaxis to prevent or reduce the frequency of episodic bleeding
        iii. Perioperative management

Criteria V — Novoseven RT

1. Hemophilia A
    a. Congenital factor VIII deficiency confirmed by blood coagulation testing; AND
    b. Confirmation that member has acquired Factor VIII inhibitors; AND
    c. Used to treat at least 1:
        i. Control and prevention of acute episodic bleeding (hemorrhage); OR
        ii. Perioperative management
2. Acquired Hemophilia
    a. Diagnosis confirmed by blood coagulation testing; AND
    b. Used to treat at least 1:
        i. Control and prevention of acute episodic bleeding (hemorrhage); OR
        ii. Perioperative management
3. Hemophilia B
    a. Factor IX deficiency confirmed by blood coagulation testing; AND
    b. Confirmation that member has acquired Factor IX inhibitors; AND
    c. Used to treat at least 1:
        i. Control and prevention of acute episodic bleeding (hemorrhage); OR
        ii. Perioperative management
4. Congenital Factor VII Deficiency
    a. Factor VII deficiency confirmed by blood coagulation testing; AND

   b. Used to treat at least 1:
      i. Control and prevention of acute episodic bleeding (hemorrhage); OR
      ii. Perioperative management
5. Glanzmann's Thrombasthenia
   a. Glanzmann's Thrombasthenia has been confirmed by blood coagulation testing; AND
   b. Used to treat at least 1:
      i. Control and prevention of acute episodic bleeding (hemorrhage); OR
      ii. Perioperative management; AND
   c. The use of platelet transfusions is known or suspected to be ineffective or contraindicated

Criteria VI — Obizur

1. Acquired Hemophilia A
   a. Factor VIII deficiency confirmed by blood coagulation testing; AND
   b. Used to treat episodic bleeding; AND
   c. Not used for congenital Hemophilia A or von Willebrand disease

Criteria VII — Tretten

1. Congenital Factor XIII A-subunit deficiency
   a. Factor XIII A-subunit deficiency confirmed by blood coagulation testing; AND
   b. Used for routine prophylaxis of bleeding

Criteria VIII — Corifact

1. Congenital Factor XIII deficiency
   a. Factor XIII deficiency confirmed by blood coagulation testing; AND
   b. Used for routine prophylactic treatment; OR
   c. Used for perioperative management of surgical bleeding

Criteria IX — Wilate

1. Von Willebrand disease (VWD)
   a. VWD confirmed by blood coagulation and von Willebrand factor testing; AND
   b. Used as treatment of spontaneous and trauma-induced bleeding episodes in at least 1:
      i. Members with severe VWD; OR

ii. Members with mild or moderate VWD in whom the use of desmopressin is known or suspected to be ineffective or contraindicated; AND

iii. Is NOT being used for routine prophylactic treatment of spontaneous bleeding episodes OR perioperative management of surgical bleeding; AND

iv. Is NOT being used for Hemophilia A

c. Used for perioperative management of bleeding

Criteria X — Hemlibra

1. Congenital Factor VIII deficiency
    a. Factor VIII deficiency confirmed by blood coagulation testing; AND
    b. Used for routine prophylactic treatment to prevent or reduce the frequency of episodic bleeding.

Criteria XI — Vonvendi

1. Von Willebrand disease (VWD)
    a. VWD confirmed by blood coagulation and von Willebrand factor testing; AND
    b. Used as treatment of spontaneous and trauma-induced bleeding episodes in at least 1:
        v. Members with severe VWD; OR
        vi. Members with mild or moderate VWD in whom the use of desmopressin is known or suspected to be ineffective or contraindicated; AND
        vii. Is NOT being used for routine prophylactic treatment of spontaneous bleeding episodes OR perioperative management of surgical bleeding; AND
        viii. Is NOT being used for Hemophilia A

Criteria XII — Coagadex

1. Hereditary Factor X deficiency
    a. Diagnosis of congenital factor X deficiency has been confirmed by blood coagulation testing; AND
        i. Used for on-demand treatment and control of bleeding episodes; OR
        ii. Used for perioperative management of surgical bleeding in patients with mild deficiency

Criteria XIII — Sevenfact (coagulation factor VIIa [recombinant]-jncw)

1. Hemophilia A (Factor VIII Deficiency)/ Hemophilia B (Factor IX Deficiency)
    a. Patient is ≥ 12 years of age; **AND**
    b. Patient has a diagnosis of Hemophilia A or B with inhibitors; **AND**
    c. Sevenfact will be used for treatment and control of bleeding episodes; **AND**
    d. Sevenfact will not be used for the treatment of congenital factor VII deficiency.

Authorization

Initial period of six months

Renewal Criteria

1. Absence of unacceptable drug-toxicity (e.g.: symptoms of allergic-anaphylactic reactions [i.e., anaphylaxis, dyspnea, rash], thromboembolic events [thromboembolism, pulmonary embolism] and development of neutralizing antibodies [inhibitors])

Limitations/Exclusions

Hemophilia medications are considered **investigational and not medically necessary** for any indications other than those listed above.

Applicable Procedure Codes

J7170	Injection, emicizumab-kxwh, 0.5 mg
J7175	Injection, factor x, (human), 1 i.u. (Coagadex)
J7179	Injection, von willebrand factor (recombinant), (vonvendi), 1 i.u. vwf:rco
J7180	Injection, factor XIII (antihemophilic factor, human), 1 IU
J7181	Injection, factor XIII A-subunit, (recombinant), per IU
J7182	Injection, factor VIII, (antihemophilic factor, recombinant), (NovoEight), per IU
J7183	Injection, von Willebrand factor complex (human), Wilate, 1 IU vWF:RCo
J7185	Injection, factor VIII (antihemophilic factor, recombinant) (XYNTHA), per IU
J7186	Injection, antihemophilic factor VIII/von Willebrand factor complex (human), per factor VIII i.u.
J7187	Injection, von Willebrand factor complex (Humate-P), per IU VWF:RCO
J7188	Injection, factor VIII (antihemophilic factor, recombinant), per IU
J7189	Factor VIIa (antihemophilic factor, recombinant), per 1 mcg
J7190	Factor VIII (antihemophilic factor, human) per IU
J7192	Factor VIII (antihemophilic factor, recombinant) per IU, not otherwise specified
J7193	Factor IX (antihemophilic factor, purified, nonrecombinant) per IU
J7194	Factor IX complex, per IU
J7195	Injection, factor IX (antihemophilic factor, recombinant) per IU, not otherwise specified
J7198	Antiinhibitor, per IU
J7199	Hemophilia clotting factor, not otherwise classified
J7200	Injection, factor IX, (antihemophilic factor, recombinant), Rixubis, per IU

J7201	Injection, factor ix, fc fusion protein, (recombinant), Alprolix, 1 i.u.
J7202	Injection, factor ix, albumin fusion protein, (recombinant), Idelvion, 1 i.u.
J7203	Injection, factor ix (antihemophilic factor, recombinant), glycopegylated, Rebinyn, 1 i.u.
J7204	Injection, factor viii, antihemophilic factor (recombinant), glycopegylated-exei, per iu (Esperoct) Effective date: 07/01/2020
J7205	Injection, factor VIII Fc fusion protein (recombinant), per IU
J7207	Injection, factor viii, (antihemophilic factor, recombinant), pegylated, 1 i.u.
J7208	Injection, factor viii (antihemophilic factor, recombinant) pegylated-aucl, Jivi, 1 i.u.
J7212	Intravenous Powder for Solution, Factor viia, (antihemophilic factor, recombinant)-jncw (sevenfact), 1 microgram
J7209	Injection, factor viii, (antihemophilic factor, recombinant), (Nuwiq), 1 i.u.
J7210	Injection, factor viii, (antihemophilic factor, recombinant), (Afstyla), 1 i.u
J7211	Injection, factor viii, (antihemophilic factor, recombinant), (Kovaltry), 1 i.u
96365	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to 1 hour
96366	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); each additional hour (List separately in addition to code for primary procedure)
96367	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); additional sequential infusion of a new drug/substance, up to 1 hour (List separately in addition to code for primary procedure)
96368	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); concurrent infusion (List separately in addition to code for primary procedure)
96369	Subcutaneous infusion for therapy or prophylaxis (specify substance or drug); initial, up to 1 hour, including pump set-up and establishment of subcutaneous infusion site(s)
96370	Subcutaneous infusion for therapy or prophylaxis (specify substance or drug); each additional hour (List separately in addition to code for primary procedure)
96371	Subcutaneous infusion for therapy or prophylaxis (specify substance or drug); additional pump set-up with establishment of new subcutaneous infusion site(s) (List separately in addition to code for primary procedure)
96372	Therapeutic, prophylactic, or diagnostic injection (specify substance or drug); subcutaneous or intramuscular
96373	Therapeutic, prophylactic, or diagnostic injection (specify substance or drug); intra-arterial
96374	Therapeutic, prophylactic, or diagnostic injection (specify substance or drug); intravenous push, single or initial substance/drug
96375	Therapeutic, prophylactic, or diagnostic injection (specify substance or drug); each additional sequential intravenous push of a new substance/drug (List separately in addition to code for primary procedure)
96376	Therapeutic, prophylactic, or diagnostic injection (specify substance or drug); each additional sequential intravenous push of the same substance/drug provided in a facility (List separately in addition to code for primary procedure)
96377	Application of on-body injector (includes cannula insertion) for timed subcutaneous injection
96379	Unlisted therapeutic, prophylactic, or diagnostic intravenous or intra-arterial injection or infusion
99601	Home infusion/specialty drug administration, per visit (up to 2 hours);
99602	Home infusion/specialty drug administration, per visit (up to 2 hours); each additional hour (List separately in addition to code for primary procedure)

Applicable ICD-10 Diagnosis Codes

D66	Hereditary factor VIII deficiency

D67	Hereditary factor IX deficiency
D68.0	Von Willebrand's disease
D68.1	Hereditary factor XI deficiency
D68.2	Hereditary deficiency of other clotting factors
D68.311	Acquired hemophilia
D68.9	Coagulation defect, unspecified
D69.1	Qualitative platelet defects
R58	Hemorrhage, not elsewhere classified

Revision History

2/20/2021	Addition of Sevenfact and its indications per FDA label. Added J7212. Removed following statement from Renewal Criteria: Member continues to meet clinical criteria above
12/28/2020	Updated Hemlibra criteria; removed the following: "Confirmation that member has Factor VIII inhibitors", in accordance with FDA labeling
06/11/2020	Added J-Code (J7204): Injection, factor viii, antihemophilic factor (recombinant), glycopegylated-exei, per iu (Esperoct) Effective date: 07/01/2020
6/03/2020	Added drug Esperoct to Criteria III in this Medical Policy
10/01/2019	Added J code J7208, for Jivi (antihemophilic factor (recombinant) pegylated-aucl Removed Related Medical Guideline for off-label use
7/10/2019	Specified Adynovate, Coagadex, Idelvion, and Vonvendi criteria
1/24/2019	addition of Jivi
7/25/2018	addition of Afstyla, Kovaltry, Rebinyn, and Hemlibra
10/13/2017	title changed from Hemophilia Infusion Therapy to Clotting Disorders Therapy. Communicated that dosing should be commensurate with drug-specific labeling and that the initial authorization period is six months.
6/16/2017	added control of acute bleeding episodes as a covered indication for Humate-P
12/3/2018	Added J7170, J7203 and removed Q9995, C9468 from Applicable Procedure Codes

References

1. Advate [package insert]. Westlake Village, CA; Baxter Healthcare. November 2016. Accessed October 20, 2017.
2. Product Information: ADYNOVATE intravenous injection, antihemophilic factor (recombinant) pegylated intravenous injection. Baxalta US Inc. (per manufacturer), Westlake Village, CA, 2017. Accessed July 10, 2019.
3. Alphanate [package insert]. Los Angeles, CA; Grifols Biologicals Inc.; March 2015. Accessed October 20, 2017.
4. AlphaNine SD [package insert]. Los Angeles, CA; Grifols Biologicals Inc.; January 2013. Accessed October 20, 2017.
5. Alprolix [package insert]. Cambridge, MA; Biogen Idec, Inc.; April 2017. Accessed October 20, 2017.
6. Bebulin [package insert]. Westlake Village, CA; Baxter Healthcare. September 2015. Accessed October 20, 2017.
7. BeneFIX [package insert]. Philadelphia, PA; Wyeth Biopharma; August 2015. Accessed October 20, 2017.
8. Product Information: COAGADEX(R) intravenous injection, coagulation factor X (human) intravenous injection. BPLUSA Inc (per FDA), Durham, NC, 2018. Accessed July 10, 2019.
9. Corifact [package insert]. Kankakee, IL; CSL Behring LLC; March 2015. Accessed October 20, 2017.
10. Eloctate [package insert]. Cambridge, MA; Biogen Idec Inc.; January 2017. Accessed October 20, 2017.
11. Fieba NF [package insert]. Westlake Village, CA; Baxter Healthcare. April 2016. Accessed October 20, 2017.
12. Helixate FS [package insert]. Kankakee, IL; CSL Behring LLC; May 2016. Accessed October 20, 2017.
13. Hemofil M [package insert]. Westlake Village, CA; Baxter Healthcare. February 2016. Accessed October 20, 2017.
14. Humate-P [package insert]. Kankakee, IL; CSL Behring LLC; June 2014. Accessed October 20, 2017.

15. Product Information: IDELVION(R) powder for intravenous injection solution, coagulation factor IX (recombinant) albumin fusion protein (rIX-FP) powder for intravenous injection solution. CSL Behring LLC, Kankakee, IL, 2016. Accessed July 10, 2019.
16. Ixinity [package insert]. Winnipeg, Manitoba, Canada. Cangene Corporation; August 2016. Accessed October 20, 2017.
17. Koate DVI [package insert]. Research Triangle Park, NC; Grifols Therapeutics Inc.; August 2012. Accessed October 20, 2017.
18. Kogenate FS [package insert]. Whippany, NJ. Bayer HealthCare LLC; August 2015. Accessed October 20, 2017.
19. MASAC Recommendations Concerning Products Licensed for the Treatment of Hemophilia and Other Bleeding Disorders. February 2016. National Hemophilia Foundation. https://www.hemophilia.org/Researchers-Healthcare-Providers/Medical-and-Scientific-Advisory-Council-MASAC/MASAC-Recommendations/MASAC-Recommendations-Concerning-Products-Licensed-for-the-Treatment-of-Hemophilia-and-Other-Bleeding-Disorders. Accessed October 20, 2017.
20. Monoclate P [package insert]. Kankakee, IL; CSL Behring LLC; February 2014. Accessed October 20, 2017.
21. Mononine [package insert]. Kankakee, IL; CSL Behring LLC; April 2016. Accessed October 20, 2017.
22. National Hemophilia Foundation. Types of Bleeding Disorders. https://www.hemophilia.org/Bleeding-Disorders/Types-of-Bleeding-Disorders. Accessed October 20, 2017.
23. Novoeight [package insert]. Bagsvaerd, Denmark; Novo Nordisk; November 2016. Accessed July October 20, 2017.
24. NovoSeven RT [package insert]. Bagsvaerd, Denmark; Novo Nordisk; March 2016. Accessed October 20, 2017.
25. NUWIQ [package insert]. Elersvagen, Sweden; Octapharma AB; September 2015. Accessed October 20, 2017.
26. Obizur [package insert]. Westlake Village, CA; Baxter Healthcare. October 2015. Accessed October 20, 2017.
27. Profilnine [package insert]. Los Angeles, CA; Grifols Biologicals Inc.; November 2013. Accessed October 20, 2017.
28. Recombinate [package insert]. Westlake Village, CA; Baxter Healthcare. August 2016. Accessed October 20, 2017.
29. Specialty-matched clinical peer review.
30. Tretten [package insert]. Bagsvaerd, Denmark; Novo Nordisk; April 2014. Accessed October 20, 2017.
31. Product Information: VONVENDI(R) lyophilized powder for intravenous injection, von Willebrand factor (recombinant) lyophilized powder for intravenous injection. Baxalta US Inc (per manufacturer), Lexington, MA, 2018. Accessed July 10, 2019.
32. Wilate [package insert]. Hoboken, NJ; Octapharma USA; August 2015. Accessed October 20, 2017.
33. Xyntha [package insert]. Philadelphia, PA; Wyeth Biopharma; December 2014. Accessed October 20, 2017.
34. KOVALTRY [package insert]. Whippany, NJ, Bayer Healthcare; March 2016. Accessed July 16, 2018.
35. AFSTYLA [package insert]. Kankakee, IL, CSL Behring LLC; September 2017. Accessed July 16, 2018.
36. REBINYN [package insert]. Plainsboro, NJ, Novo Nordisk Inc; June 2017. Accessed July 16, 2018.
37. Hemlibra [package insert]. South San Francisco, CA; Genentech; November 2017. Accessed June, 2020.
38. Jivi [package insert]. Whippany, NJ; Bayer; August 2018. Accessed January 24, 2019.
39. Esperoct [package insert]. Plainsboro, NJ: Novo Nordisk Inc; October 2019
40. Sevenfact [package insert]. Louisville, KY: HEMA Biologics, April 2020.