

## Hemophilia and Blood Factor Products

### Policy Number: C15607-A

**CRITERIA EFFECTIVE DATES:**

ORIGINAL EFFECTIVE DATE	LAST REVIEWED DATE	NEXT REVIEW DATE
02/2019	07/31/2019	07/31/2020
J CODE	TYPE OF CRITERIA	LAST P&T APPROVAL/VERSION
See products affected	RxPA	Q3 2019 20190828C15607-A

**PRODUCTS AFFECTED:**

*Plasma Factor VIII concentrates:* Hemofil M, Monoclate P, Koate DVI(J7190)

*Recombinant Factor VIII concentrates:* Advate, Helixate FS, Kogenate FS, Kovaltry, Novoeight, Nuwiq, Obizur, Recombinate, Xyntha

*Prolonged Half-Life Recombinant Factor VIII concentrates:* Adynovate, Afstyla, Eloctate

*Human Plasma-Derived Factor VIII Concentrates that Contain Von Willebrand Factor:* Alphanate, Humate P, Wilate

*Plasma Factor IX concentrates:* Alphanine SD (J1793), Mononine(J1793), Profilnine SD (J7194), Bebulin

*Recombinant Factor IX concentrates:* Rixubis, Benefix, Ixinity

*Prolonged Half-life Recombinant Factor IX concentrates:* Rebinyn, Alprolix, Idelvion

*Coagulation Factor X (Plasma-derived) agent:* Coagadex *Factor XIII Concentrate (Recombinant) agent:* Tretten

*Factor XIII Concentrate (Plasma-derived) agent:* Corifact

*Coagulation Factor VIIa (Recombinant) agent:* NovoSeven RT (J7189)

*Anti-inhibitor Coagulant Complex (Plasma-derived) agent:* FEIBA NF

*Von Willebrand factor (Recombinant) agent:* Vonvendi

*Antihemophilic Agent- Monoclonal Antibody:* Hemlibra (emicizumab)

J7193 Factor ix (antihemophilic factor, purified, non-recombinant) per i.u.

J7193 Factor ix (antihemophilic factor, purified, non-recombinant) per i.u.

J7194 Factor ix, complex, per i.u

J7194 Factor ix, complex, per i.u.

J7189 Factor viia (antihemophilic factor, recombinant), per 1 microgram

J7190 Factor viii (antihemophilic factor, human) per i.u.

J7192 Factor viii (antihemophilic factor, recombinant) per i.u., NOS

Alphanine Sd

Mononine

Profilnine

Profilnine Sd

Novoseven Rt

Koate-dvi

Recombine

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J7192 Factor viii (antihemophilic factor, recombinant) per i.u., NOS	Helixate Fs
J7192 Factor viii (antihemophilic factor, recombinant) per i.u., NOS	Kogenate Fs
J7192 Factor viii (antihemophilic factor, recombinant) per i.u., NOS	Advate
J7199 Hemophilia clotting factor, not otherwise classified	Jivi
J7203 Inj factor ix, (antihemophilic factor, recom), glycopegylated, 1 iu	Rebinyn
J7186 Inj, antihemophilic factor viii/VWF complex (human), per factor viii IU.	Alphanate/vwf
J7195 Injection, factor ix (antihemophilic factor, recombinant) per iu, NOC	Ixinity
J7195 Injection, factor ix (antihemophilic factor, recombinant) per iu, NOC	Rebinyn
J7200 Injection, factor ix, (antihemophilic factor, recombinant), rixubis, per iu	Rixubis
J7202 Injection, factor ix, albumin fusion protein, (recombinant), idelvion, 1 i.u.	Idelvion
J7201 Injection, factor ix, fc fusion protein, (recombinant), alprolix, 1 i.u.	Alprolix
J7185 Injection, factor viii (antihemophilic factor, recombinant), per i.u.	Xyntha
J7188 Injection, factor viii (antihemophilic factor, recombinant), , per i.u.	Obizur
J7205 Injection, factor viii fc fusion protein (recombinant), per iu	Eloctate
J7210 Injection, factor viii, (antihemophilic factor, recombinant), 1 i.u.	Afstyla
J7211 Injection, factor viii, (antihemophilic factor, recombinant), 1 i.u.	Kovaltry
J7182 Injection, factor viii, (antihemophilic factor, recombinant), per iu	Novoeight
J7209 Injection, factor viii, (antihemophilic factor, recombinant), 1 i.u.	Nuwiq
J7207 Injection, factor viii, (antihemophilic factor, recombinant), pegylated, 1 i.u.	Adynovate
C9141 Injection, factor viii, (antihemophilic factor, recombinant), pegylated-aucl, 1 i.u.	Jivi
J7175 Injection, factor x, (human), 1 i.u.	Coagadex
J7183 Injection, von willebrand factor complex (human), wilate, 1 i.u. wwf:rc0	Wilate
J7187 Injection, von willebrand factor complex (humate-p), per iu wwf:rc0	Humate-p
J7170 Injection, emicizumab-kxwh, 0.5 mg	Hemlibra

**DRUG CLASS:**

Antihemophilic Products

**ROUTE OF ADMINISTRATION:**

Intravenous, Subcutaneous (Hemlibra ONLY)

**PLACE OF SERVICE:**

Specialty Pharmacy

The recommendation is that medications in this policy will be for pharmacy benefit coverage and patient self-administered

**AVAILABLE DOSAGE FORMS:**

Advate SOLR 1000UNIT	Adynovate SOLR 3000UNIT	Alphanate/VWF <input type="text"/>
Advate SOLR 1500UNIT	Adynovate SOLR 500UNIT	Complex/Human SOLR
Advate SOLR 2000UNIT	Adynovate SOLR 750UNIT	1000UNIT
Advate SOLR 250UNIT	Afstyla KIT 1000UNIT	Alphanate/VWF
Advate SOLR 3000UNIT	Afstyla KIT 1500UNIT	Complex/Human SOLR
Advate SOLR 4000UNIT	Afstyla KIT 2000UNIT	1500UNIT
Advate SOLR 500UNIT	Afstyla KIT 2500UNIT	Alphanate/VWF
Adynovate SOLR 1000UNIT	Afstyla KIT 250UNIT	Complex/Human SOLR
Adynovate SOLR 1500UNIT	Afstyla KIT 3000UNIT	2000UNIT
Adynovate SOLR 2000UNIT	Afstyla KIT 500UNIT	
Adynovate SOLR 250UNIT		

Alphanate/VWF  
 Complex/Human SOLR  
 250UNIT  
 Alphanate/VWF  
 Complex/Human SOLR  
 500UNIT  
 AlphaNine SD SOLR  
 1000UNIT  
 AlphaNine SD SOLR  
 1500UNIT  
 AlphaNine SD SOLR  
 500UNIT  
 Alprolix SOLR 1000UNIT  
 Alprolix SOLR 2000UNIT  
 Alprolix SOLR 250UNIT  
 Alprolix SOLR 3000UNIT  
 Alprolix SOLR 4000UNIT  
 Alprolix SOLR 500UNIT  
 BeneFIX KIT 1000UNIT  
 BeneFIX KIT 2000UNIT  
 BeneFIX KIT 250UNIT  
 BeneFIX KIT 3000UNIT  
 BeneFIX KIT 500UNIT  
 Coagadex SOLR 250UNIT  
 Coagadex SOLR 500UNIT  
 Corifact KIT 1000-1600UNIT  
 Eloctate SOLR 1000UNIT  
 Eloctate SOLR 1500UNIT  
 Eloctate SOLR 2000UNIT  
 Eloctate SOLR 250UNIT  
 Eloctate SOLR 3000UNIT  
 Eloctate SOLR 4000UNIT  
 Eloctate SOLR 5000UNIT  
 Eloctate SOLR 500UNIT  
 Eloctate SOLR 6000UNIT  
 Eloctate SOLR 750UNIT  
 Feiba SOLR 1000UNIT  
 Feiba SOLR 2500UNIT  
 Feiba SOLR 500UNIT  
 Fibryga SOLR  
 Helixate FS KIT 1000UNIT  
 Helixate FS KIT 2000UNIT  
 Helixate FS KIT 250UNIT  
 Helixate FS KIT 3000UNIT  
 Helixate FS KIT 500UNIT  
 Hemlibra SOLN  
 105MG/0.7ML  
 Hemlibra SOLN 150MG/ML  
 Hemlibra SOLN 30MG/ML

Hemlibra SOLN 60MG/0.4ML  
 Hemofil M SOLR 1000UNIT  
 Hemofil M SOLR 1700UNIT  
 Hemofil M SOLR 250UNIT  
 Hemofil M SOLR 500UNIT  
 Humate-P SOLR 1000-  
 2400UNIT  
 Humate-P SOLR 250-  
 600UNIT  
 Humate-P SOLR 500-  
 1200UNIT  
 Idelvion SOLR 1000UNIT  
 Idelvion SOLR 2000UNIT  
 Idelvion SOLR 250UNIT  
 Idelvion SOLR 3500UNIT  
 Idelvion SOLR 500UNIT  
 Ixinity SOLR 1000UNIT  
 Ixinity SOLR 1500UNIT  
 Ixinity SOLR 2000UNIT  
 Ixinity SOLR 250UNIT  
 Ixinity SOLR 3000UNIT  
 Ixinity SOLR 500UNIT  
 Jivi SOLR 1000UNIT  
 Jivi SOLR 2000UNIT  
 Jivi SOLR 3000UNIT  
 Jivi SOLR 500UNIT  
 Kcentra KIT 1000UNIT  
 Kcentra KIT 500UNIT  
 Koate SOLR 1000UNIT  
 Koate SOLR 250UNIT  
 Koate SOLR 500UNIT  
 Koate-DVI SOLR 1000UNIT  
 Koate-DVI SOLR 250UNIT  
 Koate-DVI SOLR 500UNIT  
 Kogenate FS KIT 1000UNIT  
 Kogenate FS KIT 2000UNIT  
 Kogenate FS KIT 250UNIT  
 Kogenate FS KIT 3000UNIT  
 Kogenate FS KIT 500UNIT  
 Kovaltry SOLR 1000UNIT  
 Kovaltry SOLR 2000UNIT  
 Kovaltry SOLR 250UNIT  
 Kovaltry SOLR 3000UNIT  
 Kovaltry SOLR 500UNIT  
 Mononine SOLR 1000UNIT  
 Novoeight SOLR 1000UNIT  
 Novoeight SOLR 1500UNIT  
 Novoeight SOLR 2000UNIT  
 Novoeight SOLR 250UNIT

Novoeight SOLR 3000UNIT  
 Novoeight SOLR 500UNIT  
 NovoSeven RT SOLR 1MG  
 NovoSeven RT SOLR 2MG  
 NovoSeven RT SOLR 5MG  
 NovoSeven RT SOLR 8MG  
 Nuwiq KIT 1000UNIT  
 Nuwiq KIT 2000UNIT  
 Nuwiq KIT 2500UNIT  
 Nuwiq KIT 250UNIT  
 Nuwiq KIT 3000UNIT  
 Nuwiq KIT 4000UNIT  
 Nuwiq KIT 500UNIT  
 Nuwiq SOLR 1000UNIT  
 Nuwiq SOLR 2000UNIT  
 Nuwiq SOLR 2500UNIT  
 Nuwiq SOLR 250UNIT  
 Nuwiq SOLR 3000UNIT  
 Nuwiq SOLR 4000UNIT  
 Nuwiq SOLR 500UNIT  
 Obizur SOLR 500UNIT  
 Profilnine SD SOLR  
 1000UNIT  
 Profilnine SD SOLR  
 1500UNIT  
 Profilnine SD SOLR  
 500UNIT  
 Profilnine SOLR 1000UNIT  
 Profilnine SOLR 1500UNIT  
 Profilnine SOLR 500UNIT  
 Rebinyn SOLR 1000UNIT  
 Rebinyn SOLR 2000UNIT  
 Rebinyn SOLR 500UNIT  
 Recombinate SOLR 1241-  
 1800UNIT  
 Recombinate SOLR 1801-  
 2400UNIT  
 Recombinate SOLR 220-  
 400UNIT  
 Recombinate SOLR 401-  
 800UNIT  
 Recombinate SOLR 801-  
 1240UNIT  
 RiaSTAP SOLR  
 Rixubis SOLR 1000UNIT  
 Rixubis SOLR 2000UNIT  
 Rixubis SOLR 250UNIT  
 Rixubis SOLR 3000UNIT  
 Rixubis SOLR 500UNIT



Tretten SOLR 2000-3125UNIT  
 Vonvendi SOLR 1300UNIT  
 Vonvendi SOLR 650UNIT  
 Wilate KIT 1000-1000UNIT  
 Wilate KIT 500-500UNIT  
 Xyntha KIT 1000UNIT

Xyntha KIT 2000UNIT  
 Xyntha KIT 250UNIT  
 Xyntha KIT 500UNIT  
 Xyntha Solofuse KIT 1000UNIT  
 Xyntha Solofuse KIT 2000UNIT

Xyntha Solofuse KIT 250UNIT  
 Xyntha Solofuse KIT 3000UNIT  
 Xyntha Solofuse KIT 500UNIT

**FDA-APPROVED USES:**

control and prevention of Hemophilia A hemorrhage, control and prevention of Hemophilia B hemorrhage, hemorrhage in von Willebrand disorder, treatment of hemorrhage in congenital fibrinogen deficiency, acquired factor VIII deficiency disease, congenital factor VII deficiency, Glanzmann's thrombasthenia, hemophilia, with inhibitors to Factor VIII or Factor IX  
 OBIZUR ONLY: routine prophylaxis in Hemophilia A, surgical procedure prophylaxis in Hemophilia A, control of hemorrhage in Hemophilia A

**COMPENDIAL APPROVED OFF-LABELED USES:** None

**COVERAGE CRITERIA: INITIAL AUTHORIZATION**

**DIAGNOSIS:** Indicated for use in children and adults with hemophilia A (congenital factor VIII deficiency)

**REQUIRED MEDICAL INFORMATION:**

A. FOR ALL INDICATIONS:

1. Documentation of diagnosis and the requested dose and frequency is in accordance with FDA-approved labeling, nationally recognized compendia, and/or evidence-based practice guidelines AND
2. Documentation of the prescription drug, dose and directions along with requested assay and quantity dated within the last 12 months AND
3. Documentation prescriber has verified that the number of on-hand doses (prn doses) in members home doses not exceed three doses AND
4. FOR HEMLIBRA ONLY: Documentation patient has developed high-titer factor VIII inhibitors ( > 5 Bethesda units [BU]) AND Prescribed for the prevention of bleeding episodes (i.e., routine prophylaxis)

**DURATION OF APPROVAL:**

Initial authorization: 3 months, Continuation of therapy: 12 months

**QUANTITY:** No requirements

**PRESCRIBER REQUIREMENTS:**

Prescribed by or in consultation with a hematologist or specialist at a Hemophilia treatment center ([https://www2a.cdc.gov/ncbddd/htcweb/Dir\\_Report/Dir\\_Search.asp](https://www2a.cdc.gov/ncbddd/htcweb/Dir_Report/Dir_Search.asp))- Directory

**AGE RESTRICTIONS:**

None

**GENDER:**

Male and female

**CONTINUATION OF THERAPY:****A. FOR ALL INDICATIONS:**

1. Documentation of the current number of on-hand doses and number of bleeding episodes since the previous authorization  
AND
2. For prophylactic dosing, coverage will not be granted for additional doses if greater than a 7-day supply is on hand unless on-demand dosing is medically necessary  
AND
3. If inhibitors are present, Bethesda assay titers are required  
AND
4. For treatment with ITI, continued therapy is no longer considered medically necessary if either of the following criteria are met: Inhibitor levels become undetectable (negative Bethesda assay) and Recovery of Factor VIII levels after infusion are normal (defined as at least 66% of expected level and a half-life of >6 hours are considered sufficient normal pharmacokinetic responses to characterize a complete tolerance

**CONTRAINDICATIONS/EXCLUSIONS/DISCONTINUATION:**

All other uses of Hemophilia and Blood Factor Products are considered experimental/investigational and therefore, will follow Molina's Off-Label policy

**OTHER SPECIAL CONSIDERATIONS:**

None

**BACKGROUND:**

Hemophilia and von Willebrand's disease are the most common congenital bleeding disorders. The two main types of hemophilia are A and B. Hemophilia A (classic hemophilia) has low levels of clotting factor VIII, or antihemophilic factor (AHF). Hemophilia B (Christmas disease) has low levels of clotting factor IX. AHF is an endogenous glycoprotein necessary for blood clotting and hemostasis. It is a cofactor that is necessary for factor IX to activate factor X in the intrinsic pathway.

The main treatment for hemophilia is replacement of clotting factor VIII (for hemophilia A) or clotting factor IX (for hemophilia B). Administration of clotting factors is indicated for hemophilia when a bleeding episode arises (demand treatment) or when bleeding is anticipated or likely (prophylactic treatment).

Hemophilia A and B are classified as mild, moderate or severe, depending on the amount of clotting factor VIII or IX in the blood.

Mild hemophilia: 5 – 40 percent of normal clotting factor

Moderate hemophilia: 1 – 5 percent of normal clotting factor

Severe hemophilia: Less than 1 percent of normal clotting factor

**APPENDIX: None**

1. Advate (antihemophilic factor [recombinant]) [prescribing information]. Westlake Village, CA: Baxalta US Inc; November 2016.
2. Afstyla (antihemophilic factor [recombinant]) [prescribing information]. Kankakee, IL: CSL Behring LLC; September 2017.
3. Adynovate [package insert]. Westlake Village, CA: Baxalta US Inc.; December 2016.
4. Eloctate [package insert]. Cambridge, MA: Biogen Idec Inc.; January 2017.
5. Helixate FS [package insert]. Whippany, NJ: Bayer HealthCare LLC; May 2016.
6. Kogenate FS [package insert]. Whippany, NJ: Bayer HealthCare LLC; May 2016.
7. Kogenate FS with BIO-SET [package insert]. Whippany, NJ: Bayer HealthCare LLC; May 2016.
8. Kogenate FS with Vial Adapter [package insert]. Whippany, NJ: Bayer HealthCare LLC; May 2016.
9. Hemlibra (emicizumab-kxwh) [prescribing information]. South San Francisco, CA: Genentech Inc; October 2018.
10. Kovaltry [package insert]. Whippany, NJ: Bayer Healthcare LLC; March 2016.
11. Novoeight [package insert]. Plainsboro, NJ: Novo Nordisk Inc., November 2016.
12. Nuwiq [package insert]. Peyvandi F, Garagiola I, Young G. The past and future of haemophilia: diagnosis, treatments, and its complications. *Lancet* 2016; 388:187.
13. Xyntha (antihemophilic factor [recombinant]) [prescribing information]. Philadelphia, PA: Wyeth Pharmaceuticals Inc; October 2014.
14. BeneFix (coagulation factor IX [recombinant]) [prescribing information]. Philadelphia, PA: Wyeth Pharmaceuticals; June 2017
15. Rixubis (coagulation factor IX [recombinant]) [prescribing information]. Westlake Village, CA: Baxalta US Inc; March 2016
16. Zonovate (antihemophilic factor [recombinant]) [product monograph]. Mississauga, Ontario, Canada: Novo Nordisk Canada Inc; May 2017.
17. Recombinate (antihemophilic factor [recombinant]) [prescribing information]. Westlake Village, CA: Baxalta US; March 2017
18. Eloctate (antihemophilic factor [recombinant]) [prescribing information]. Cambridge, MA: Biogen Idec; January 2017
19. Alprolix (coagulation factor IX [recombinant]) [prescribing information]. Cambridge, MA: Biogen Idec; June 2018.
20. NovoSeven RT (factor VIIa) [prescribing information]. Plainsboro, NJ: Novo Nordisk; October 2017.
20. Hemlibra (emicizumab-kxwh) [prescribing information]. South San Francisco, CA: Genentech Inc; October 2018.
21. Berntorp E, Shapiro AD. Modern haemophilia care. *Lancet* 2012; 379:1447.
21. Oldenburg J. Optimal treatment strategies for hemophilia: achievements and limitations of current prophylactic regimens. *Blood* 2015; 125:2038. <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 on December 21, 2016).
22. Srivastava A, Brewer AK, Mauser-Bunschoten EP, et al. Guidelines for the management of hemophilia. *Haemophilia* 2013; 19:e1.