Medical Policy

Healthcare Services Department

Policy Name	Policy Number	Scope
Agents for Hemophilia A and Von Willebrand Disease	MP-RX-FP-02-23	☒ MMM MA ☒ MMM Multihealth

Service Category

☐ Anesthesia
☐ Surgery
☐ Radiology Procedures
☐ Pathology and Laboratory Procedures
☐ Medicine Services and Procedures
☐ Evaluation and Management Services
☐ DME/Prosthetics or Supplies
☒ Part B Drugs

Service Description

This document addresses the use of Antihemophilic Factor VIII Human [Koate, Hemofil-M], Antihemophilic Factor (Recombinant) Plasma/Albumin-Free [Advate, Afstyla, Kogenate FS, Kovaltry, Novoeight, Nuwiq, Recombinate, Xyntha/Xyntha Solofuse], Antihemophilic Factor (factor VIII) – Long acting [– Adynovate, Jivi, Eloctate, Esperoct, Altuviiio], Anti-hemophilic bispecific factor (Factor IXa- and Factor X-)[Hemlibra (emicizumab-kxwh)], Anti-hemophilic Factor VIII (Recombinant), Porcine Sequence [Obizur], Anti-hemophilic Factor VIII/von Willebrand Factor Complex [Alphanate, Humate-P, Wilate], von Willebrand factor, Recombinant [Vonvendi] , drugs approved by the Food and Drug Administration (FDA) for the treatment of Hemophilia A and von Willebrand Disease.

Background Information
Agents in this document are used for hereditary or congenital hemophilia A, also called factor VIII (FVIII) deficiency or classic hemophilia. Acquired hemophilia A (also called acquired factor VIII deficiency) is a rare autoimmune disorder, and not a congenital disease. This document does not address fibrin products, fibrin sealants and blood products provided by blood banks. Bypassing agents (i.e., NovoSeven RT, SevenFact, and FEIBA) for those who develop antibodies or inhibitors to factor products, and Stimate (desmopressin acetate) intranasal spray are discussed in separate documents.

Factor replacement treatments can be created from blood products (human plasma-derived) and others that are manufactured (recombinant). Replacement therapy may be given on a routine, preventive basis which is also called prophylactic therapy. The infusion of factor replacements given to stop a bleeding episode is called on-demand or episodic therapy. While the World Federation of Hemophilia (WFH) (Srivastava 2020) does not place a preference between plasma-derived and recombinant products, the U.S. National Hemophilia Federation (NHF 2020) recommends recombinant over plasma-derived due the possibility of virus transmission. WFH states that the choice between the two classes of products must be made according to the availability, cost, and patient preferences.

Products in this document include:

- Anti-hemophilic factor (factor VIII) Human plasma derived: Hemofil-M, Koate-DVI
- Anti-hemophilic factor (factor VIII) Recombinant: Advate, Afstyla, Kogenate FS, Kovaltry, Novoeight, Nuwiq, Recombinate, Xyntha/Xyntha Solofuse
- Anti-hemophilic factor (factor VIII) – Long acting
  o Recombinant, Pegylated – Adynovate,

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o Recombinant, Pegylated damactocog alfa pegol – Jivi
o Recombinant Fc Fusion Protein–Eloctate
o Recombinant, Glycopegylated– Esperoct o Recombinant Anti-hemophilic factor Fc-VWF-XTEN Fusion Protein-ehtl – Altuviiio

- Anti-hemophilic bispecific factor (Factor IXa- and Factor X-): Hemlibra (emicizumab-kxwh)
- Anti-hemophilic Factor VIII (Recombinant), Porcine Sequence: Obizur
- Anti-hemophilic Factor VIII/von Willebrand Factor Complex : Alphanate, Humate-P, Wilate
- von Willebrand factor, Recombinant: Vonvendi

Hereditary hemophilia A is the most common type of hemophilia. Although it is usually inherited, about one third of cases are caused by spontaneous mutations. Hemophilia A is related to mutations in the gene coding for coagulation Factor VIII, and it is four times more common than hemophilia B (CDC 2014), the second most common hemophilia type.

The U.S. National Hemophilia Foundation (NHF) and the World Federation of Hemophilia (Srivastava 2020) both note there is a relationship of bleeding severity to the clotting factor level. Both entities list “severe” hemophilia as a clotting factor level < 1 IU/dl or < 1% of normal. A “mild” bleeding severity is identified as a clotting factor level of 5-40 IU/dl or 5 to < 40% of normal. A bleeding episode for individuals with mild risk includes severe bleeding with major trauma or surgery. Individuals with 1-5 IU/dl or 1-5% of normal are considered “moderate” risk for occasional spontaneous bleeding and prolonged bleeding with minor trauma or surgery (Srivastava 2020).

Hemophilia severity:

A. Severe hemophilia – Severe hemophilia is defined as < 1 percent factor activity, which corresponds to < 1 IU/dL.
B. Moderate hemophilia – Moderate hemophilia is defined as a factor activity level ≥ 1 percent of normal and < 5 percent of normal, corresponding to ≥ 1 and < 5 IU/dL.
C. Mild hemophilia – Mild hemophilia is defined as a factor activity level ≥ 5 percent of normal and < 40 percent of normal (≥ 5 and < 40 IU/dL).

World Federation of Hemophilia 2020 Guidelines for treatment of hemophilia state that prophylaxis prevents bleeding and joint destruction, and that prophylaxis should enable those with hemophilia to lead healthy and active lives. Moreover, the updated 2020 guidelines proposes that the definition of prophylaxis be based on outcomes rather than doses or timing of initiation, and treatment regimens that take into account the hemophilic phenotype of the individual in addition to factor levels. However, more studies are needed to determine if all individuals should remain on therapy as adults (that is, those with severe hemophilia vs. moderate or mild). The WFH 2020 guidelines have been endorsed by several societies worldwide, including the U.S. NHF. Short-term prophylaxis (of 4 to 8 weeks) may interrupt the bleeding cycle and benefit individuals with repeated bleeding into target joints. Prophylaxis does not reverse existing joint damage but reduces bleeding and may slow progression of joint damage. Prophylactic clotting factor administration is recommended prior to the individual engaging in activities with higher risk of injury. Randomized trials of

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prophylactic therapy of hemophilia have demonstrated a decreased incidence of arthropathy (Gringeri, 2011; Manco-Johnson, 2007).

Von Willebrand disease (VWD) is the most common inherited bleeding disorder. It is caused by missing for defective von Willebrand factor (VWF), a clotting protein. Unlike hemophilia, which very rarely occurs in females, von Willebrand disease can occur equally in men and women. There are three main types of hereditary VWD – type 1 (most common), type 2, and type 3. The types are classified by their level of VWF in the blood or by the presence or behavior of the VWF chains. VWD can be caused by an autoimmune disorder or as a result of certain medications. This is called acquired von Willebrand disease. Those with VWD can experience frequent nosebleeds, easy bruising, and excessive bleeding after during and after surgical procedures. Women may have heavy and long lasting menstrual periods and hemorrhaging after childbirth (NHF).

Clinical Criteria:

Anti-hemophilic factor (factor VIII) Human plasma derived: Koate-DVI, Hemofil M

Initial requests for Hemofil M or Koate/Koate-DVI (Factor VIII, human plasma-derived) may be approved if the following criteria are met:

I. Individual has a diagnosis of hemophilia A (also called factor VIII deficiency or classic hemophilia); **AND**
II. Individual is using for the treatment of bleeding episodes;

OR
III. Individual has a diagnosis of hemophilia A (also called factor VIII deficiency or classic hemophilia); **AND**
IV. Individual is using as routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
V. Individual has a diagnosis of severe hemophilia A (defined as less than 1 International Unit per deciliter [1IU/dL] or 1% endogenous Factor VIII) (NHF, Srivastava 2020);

OR
VI. Individual has a diagnosis of hemophilia A (also called factor VIII deficiency or classic hemophilia); **AND**
VII. Individual is using as routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
VIII. Individual has a diagnosis of mild to moderate hemophilia A (defined as endogenous Factor VIII less than 40 IU/dL [less than 40%], but greater than or equal to 1 IU) (NHF, Srivastava 2020); **AND**
IX. Individual has one of the following (NHF, Srivastava 2020):

A. One or more episodes of spontaneous bleeding into joint; **OR**
B. One or more episodes of severe, life-threatening, or spontaneous bleeding as determined by the prescriber; **OR**
C. Severe phenotype hemophilia determined by the individual's risk factors that increase the risk of a clinically significant bleed, including but not limited to, participation in activities likely to cause

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injury/trauma, procoagulant and anticoagulant protein levels, comorbid conditions affecting functional ability and physical coordination, or history of a clinically significant bleed.

Initial requests for Koate/Koate-DVI (Factor VIII, human plasma-derived) may be approved if the following criteria are met:

I. Individual has a diagnosis of hemophilia A (also called factor VIII deficiency or classic hemophilia); AND
II. Individual is using for peri-procedural management for surgical, invasive or interventional radiology procedures.

Continuation requests for Hemofil M or Koate/Koate-DVI (Factor VIII, human plasma-derived) may be approved if the following criteria are met:

I. Individual has had a positive therapeutic response to treatment (for example, reduction in frequency and/or severity of bleeding episodes).

Hemofil M, Koate/Koate-DVI (Factor VIII, human plasma-derived) **may not be approved** for the following:
I. Individual is using for the treatment of von Willebrand disease (VWD); OR
II. When the above criteria are not met and for all other indications.

Anti-hemophilic factor (factor VIII) Recombinant: Advate, Afstyla, Kogenate FS, Kovaltry, Novoeight, Nuwiq, Recombinate, Xyntha/Xyntha Solofuse

Initial requests for Advate, Afstyla, Kogenate FS, Kovaltry, Novoeight, Nuwiq, Recombinate, or Xyntha/Xyntha Solofuse (Factor VIII recombinant) may be approved if the following criteria are met:

I. Individual has a diagnosis of hemophilia A (also called factor VIII deficiency or classic hemophilia); **AND**
II. Individual is using for one of the following:
A. Treatment of bleeding episodes; **OR**
B. Peri-procedural management for surgical, invasive or interventional radiology procedures;

OR
III. Individual has a diagnosis of von Willebrand disease; **AND**
IV. Individual is using for the treatment of bleeding episodes; **AND**
V. Individual is using in combination with Vonvendi (recombinant von Willebrand factor complex); **AND**
VI. Individual has a baseline factor VIII level less than 40 IU/dL [less than 40%] or are unknown (Vonvendi 2018);

OR
VII. Individual has a diagnosis of von Willebrand disease; **AND**
VIII. Individual is using for peri-procedural management for surgical, invasive or interventional radiology procedures; **AND**

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IX. Individual is using in combination with Vonvendi (recombinant von Willebrand factor complex); **AND**
X. Individual has a baseline factor VIII level less than 30 IU/dL [less than 30%] or are unknown (Vonvendi 2018).

Initial requests for Advate, Afstyla, Kogenate FS, Kovaltry, Novoeight, Nuwiq, or Xyntha/Xyntha Solofuse (Factor VIII recombinant) may be approved if the following criteria are met:

I. Individual is using as routine prophylaxis to prevent or reduce the frequency of bleeding episodes; AND
II. Individual has a diagnosis of severe hemophilia A (defined as less than 1 International Unit per deciliter [1IU/dL] or 1% endogenous Factor VIII) (NHF, Srivastava 2020);

OR
III. Individual has a diagnosis of mild to moderate hemophilia A (defined as endogenous Factor VIII less than 40 IU/dL [less than 40%], but greater than or equal to 1 IU) (NHF, Srivastava 2020); **AND**
IV. Individual is using as routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
V. Individual has one of the following:
- A. One or more episodes of spontaneous bleeding into joint; **OR**
- B. One or more episodes of severe, life-threatening, spontaneous bleeding as determined by the prescriber; **OR**
- C. Severe phenotype hemophilia determined by the individual's risk factors that increase the risk of a clinically significant bleed, including but not limited to, participation in activities likely to cause injury/trauma, procoagulant and anticoagulant protein levels, comorbid conditions affecting functional ability and physical coordination, or history of a clinically significant bleed.

Initial requests for Kogenate FS (Factor VIII recombinant) may be approved if the following criteria are met:

I. Individual is 16 years of age or younger; **AND**
II. Individual has a diagnosis of hemophilia A (also called factor VIII deficiency or classic hemophilia); **AND**
III. Individual is using as routine prophylaxis to reduce the risk of joint damage in those without pre-existing joint damage.

Initial requests for Recombinate (Factor VIII recombinant) may be approved if the following criteria are met:

I. Individual is using for the treatment of acquired Factor VIII inhibitors not exceeding 10 Bethesda Unit (BU) per milliliter (mL).

Continuation requests for Advate, Afstyla, Kogenate FS, Kovaltry, Novoeight, Nuwiq, Recombinate, or Xyntha/Xyntha Solofuse (Factor VIII recombinant) may be approved if the following criteria are met:

I. Individual has had a positive therapeutic response to treatment (for example, reduction in frequency and/or severity of bleeding episodes).

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Advate, Afstyla, Kogenate FS, Kovaltry, Novoeight, Nuwiq, Recombinate, or Xyntha/Xyntha Solofuse (Factor VIII recombinant) **may not be approved** for the following:

I. Individual is using as monotherapy for the maintenance treatment of von Willebrand disease; OR
II. When the above criteria are not met and for all other indications.

Anti-hemophilic factor (factor VIII) – Long acting: Adynovate (Factor VIII Long-Acting Recombinant, pegylated), Jivi (Factor VIII Recombinant, PEGylated damactocog alfa pegol), Eloctate (Factor VIII Recombinant Anti-hemophilic Factor Fc Fusion Protein), Esperoct (Factor VIII Recombinant, glycopegylated), or Factor VIII Recombinant Antihemophilic Factor FC-VWF-XTEN Fusion Protein (Altuviiio).

Initial requests for Adynovate (Factor VIII Long-Acting Recombinant, pegylated), Jivi (Factor VIII Recombinant PEGylated damactocog alfa pegol), Eloctate (Factor VIII Recombinant Anti-hemophilic Factor Fc Fusion Protein), Esperoct (Factor VIII Recombinate, glycopegylated), or Altuviiio (Factor VIII Recombinant Antihemophilic Factor FC-VWF-XTEN Fusion Protein) may be approved if the following criteria are met:
I. Individual has a diagnosis of severe hemophilia A (defined as less than 1 International Unit per deciliter [1IU/dL] endogenous Factor VIII) (NHF, Srivastava 2020); **AND**
II. Individual is using for one of the following:
A. Treatment of acute bleeding episodes; **OR**
B. Peri-procedural management for surgical, invasive or interventional radiology procedures; **OR**
C. Routine prophylaxis to prevent or reduce the frequency of bleeding episodes;
AND
III. If using Jivi, individual is 12 years of age or older and has been previously treated with factor VIII;

OR
IV. Individual has a diagnosis of mild to moderate hemophilia A (defined as endogenous Factor VIII less than 40 IU/dL [less than 40%], but greater than or equal to 1 IU) (NHF, Srivastava 2020); **AND**
V. Individual is using for one of the following:
A. Treatment of acute bleeding episodes; **OR**
B. Peri-procedural management for surgical, invasive or interventional radiology procedures; **OR**
C. Routine prophylaxis to prevent or reduce the frequency of bleeding episodes when one of the following:
AND
1. Individual has had one or more episodes of spontaneous bleeding into joint; **OR**
2. Individual has had one or more episodes of severe, life-threatening, or spontaneous bleeding as determined by the prescriber; **OR**
3. Severe phenotype hemophilia determined by the individual's risk factors that increase the risk of a clinically significant bleed, including but not limited to, participation in activities likely to cause injury/trauma, procoagulant and anticoagulant protein levels, comorbid conditions affecting functional ability and physical coordination, or history of a clinically significant bleed;
AND
VI. If using Jivi, individual is 12 years of age or older and has been previously treated with factor VIII.

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Continuation requests for Adynovate (Factor VIII Long-Acting Recombinant, pegylated), Jivi (Factor VIII Recombinant PEGylated damactocog alfa pegol), Eloctate (Factor VIII Recombinant Anti-hemophilic Factor Fc Fusion Protein), Esperoct (Factor VIII Recombinant, glycopegylated), or Altuviiio (Factor VIII Recombinant Antihemophilic Factor FC-VWF-XTEN Fusion Protein) may be approved if the following criteria are met:

I. Individual has had a positive therapeutic response to treatment (for example, reduction in frequency and/or severity of bleeding episodes).

Adynovate (Factor VIII Long-Acting Recombinant, pegylated), Jivi (Factor VIII Recombinant PEGylated damactocog alfa pegol), Eloctate (Factor VIII Recombinant Anti-hemophilic Factor Fc Fusion Protein), Esperoct (Factor VIII Recombinant, glycopegylated), or Altuviiio (Factor VIII Recombinant Antihemophilic Factor FC-VWF-XTEN Fusion Protein) **may not be approved** for the following:

I. Individual is using for the treatment of von Willebrand disease; **OR**
II. When the above criteria are not met and for all other indications.

Hemlibra (emicizumab-kxwh) - Anti-hemophilic bispecific factor --- Factor IXa and Factor X

Initial requests for Hemlibra (emicizumab-kxwh) may be approved if the following criteria are met:

I. Individual has a diagnosis of severe hemophilia A (defined as less than 1 International Unit per deciliter [1IU/dL] endogenous Factor VIII) (NHF, Srivastava 2020); **AND**
II. Individual is using for routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
III. Individual has one of the following:
A. If switching from factor VIII agents, then individual will discontinue factor VIII agents being used for routine prophylaxis after the first week of Hemlibra initiation; **OR**
B. If switching from bypassing agents (i.e., NovoSeven RT, SevenFact, FEIBA), then individual will discontinue bypassing agents being used for routine prophylaxis after 24 hours of Hemlibra initiation;

OR
IV. Individual has a diagnosis of mild to moderate hemophilia A (defined as endogenous Factor VIII less than 40 IU/dL [less than 40%], but greater than or equal to 1 IU) (NHF, Srivastava 2020); **AND**
V. Individual is using for routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
VI. Individual has one of the following:
A. One or more episodes of spontaneous bleeding into joint; **OR**
B. One or more episodes of severe, life-threatening, or spontaneous bleeding as determined by the prescriber; **OR**
C. Severe phenotype hemophilia determined by the individual's risk factors that increase the risk of a clinically significant bleed, including but not limited to, participation in activities likely to cause injury/trauma,

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procoagulant and anticoagulant protein levels, comorbid conditions affecting functional ability and physical coordination, or history of a clinically significant bleed; **AND**
VII. Individual has one of the following:
A. If switching from factor VIII agents, then individual will discontinue factor VIII agents being used for routine prophylaxis after the first week of Hemlibra initiation; **OR**
B. If switching from bypassing agents, (i.e., NovoSeven RT, SevenFact, FEIBA), then individual will discontinue bypassing agents being used for routine prophylaxis after 24 hours of Hemlibra initiation.

Continuation requests for Hemlibra (emicizumab-kxwh) may be approved if the following criteria are met:

I. Individual has had a positive therapeutic response to treatment (for example, reduction in frequency and/or severity of bleeding episodes).

Hemlibra (emicizumab) **may not be approved** when the above criteria are not met and for all other indications.

Obizur (Factor VIII Recombinant, Porcine Sequence)

Initial requests for Obizur (Recombinant, Porcine Sequence) may be approved if the following criteria are met:

I. Individual is 18 years of age or older; **AND**
II. Individual has a diagnosis of acquired hemophilia A; **AND**
III. Individual has baseline anti-porcine Factor VIII inhibitor titer less than or equal to 20 BU/mL; **AND**
IV. Individual is using for the treatment of bleeding episodes.

Continuation requests for Obizur (Recombinant, Porcine Sequence) may be approved if the following criteria are met:

I. Individual has had a positive therapeutic response to treatment (for example, reduction in frequency and/or severity of bleeding episodes).

Obizur (Recombinant, Porcine Sequence**) may not be approved** for the following:

I. Individual has a diagnosis of congenital hemophilia A with Factor VIII deficiency; OR
II. Individual has a diagnosis of von Willebrand disease; OR
III. When the above criteria are not met and for all other indications.

Alphanate, Humate-P, Wilate (Anti-hemophilic Factor VIII/von Willebrand Factor Complex, Human)

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Initial requests for Alphanate, Humate-P, or Wilate (Anti-hemophilic Factor VIII/von Willebrand Factor Complex, Human) may be approved if the following criteria are met:

I. Individual has a diagnosis of hemophilia A (also called factor VIII deficiency or classic hemophilia); **AND**
II. Individual is using for the treatment of bleeding episodes;

OR
III. Individual has a diagnosis of severe hemophilia A (defined as less than 1 International Unit per deciliter [1IU/dL] endogenous Factor VIII) (NHF, Srivastava 2020); **AND**
IV. Individual is using for routine prophylaxis to prevent or reduce the frequency of bleeding episodes;

OR
V. Individual has a diagnosis of mild to moderate hemophilia A (defined as endogenous Factor VIII less than 40 IU/dL [less than 40%], but greater than or equal to 1 IU) (NHF, Srivastava 2020); **AND**
VI. Individual is using for routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
VII. Individual has one of the following:
A. Individual has had one or more episodes of spontaneous bleeding into joint; **OR**
B. Individual has had one or more episodes of severe, life-threatening, or spontaneous bleeding as determined by the prescriber; **OR**
C. Severe phenotype hemophilia determined by the individual's risk factors that increase the risk of a clinically significant bleed, including but not limited to, participation in activities likely to cause injury/trauma, procoagulant and anticoagulant protein levels, comorbid conditions affecting functional ability and physical coordination, or history of a clinically significant bleed.

Initial requests for Alphanate (Anti-hemophilic Factor VIII/von Willebrand Factor Complex, Human) may be approved if the following criteria are met:

I. Individual has a diagnosis with acquired Factor VIII deficiency; **AND**
II. Individual is using for the control and prevention of bleeding episodes.

Initial requests for Alphanate, Humate-P, Wilate (Anti-hemophilic Factor VIII/von Willebrand Factor Complex, Human) may be approved if the following criteria are met:

I. Individual has a diagnosis of severe von Willebrand disease; **OR**
II. Individual has a diagnosis of mild to moderate von Willebrand disease and use of desmopressin is known or suspected to be inadequate; **AND**
III. Individual is using for one of the following:
A. The treatment of spontaneous or trauma-induced bleeding episodes; **OR**
B. Peri-procedural management for surgical, invasive or interventional radiology procedures.

Continuation requests for Alphanate, Humate-P, or Wilate (Anti-hemophilic Factor VIII/von Willebrand Factor Complex, Human) may be approved if the following criteria are met:

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I. Individual has had a positive therapeutic response to treatment (for example, reduction in frequency and/or severity of bleeding episodes).

Alphanate (Anti-hemophilic Factor VIII/von Willebrand Factor Complex, Human) **may not be approved** for the following:
I. Individual has a diagnosis for severe (type 3) von Willebrand Disease; **AND**
II. Individual is undergoing major surgery;
OR
III. Individual is using for prophylaxis of spontaneous bleeding episodes in von Willebrand disease.

Humate-P and Wilate (Anti-hemophilic Factor VIII/von Willebrand Factor Complex, Human) **may not be approved** for the following:

I. Individual is using for prophylaxis of spontaneous bleeding episodes in von Willebrand disease.

Alphanate, Humate-P, Wilate (Anti-hemophilic Factor VIII/von Willebrand Factor Complex, Human) **may not be approved** when the above criteria are not met and for all other indications.

Vonvendi (Recombinant von Willebrand Factor Complex)

Initial requests for Vonvendi (Recombinant von Willebrand Factor Complex) may be approved if the following criteria are met:

I. Individual is 18 years of age or older; **AND**
II. Individual is using for one of the following:
A. Individual has a diagnosis of severe von Willebrand disease; **OR**
B. Individual has a diagnosis of mild to moderate von Willebrand disease and use of desmopressin is known or suspected to be inadequate;
AND

III. Individual is using for one of the following:
A. Individual is using to treat spontaneous or trauma-induced bleeding episodes, or for peri-procedural management for surgical, invasive or interventional radiology procedures; **OR**
B. Individual is using as routine prophylaxis to prevent or reduce the frequency of bleeding episodes receiving on-demand therapy.

Continuation requests for Vonvendi (Recombinant von Willebrand Factor Complex) may be approved if the following criteria are met:

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I. Individual has had a positive therapeutic response to treatment (for example, reduction in frequency and/or severity of bleeding episodes).

Vonvendi (Recombinant von Willebrand Factor Complex) **may not be approved** when the above criteria are not met and for all other indications.

Applicable Codes

The following list(s) of procedure and/or diagnosis codes is provided for reference purposes only and may not be all inclusive. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement policy. Benefit coverage for health services is determined by the member specific benefit plan document and applicable laws that may require coverage for a specific service. The inclusion of a code does not imply any right to reimbursement or guarantee claim payment. Other Policies and Guidelines may apply.

Anti-hemophilic factor (factor VIII) Human plasma derived: Koate-DVI, Hemofil M

HCPCS	Description
J7190	Factor VIII Anti-hemophilic factor, human, per IU [Hemofil M, Koate DVI,]

ICD-10	Description
D66	Hereditary factor VIII deficiency [hemophilia A]
D68.00-D68.09	Von Willebrand's disease
D68.311	Acquired hemophilia
D68.318	Other hemorrhagic disorder due to intrinsic circulating anticoagulants, antibodies, or inhibitors
D68.4	Acquired coagulation factor deficiency
Z29.8	Encounter for other specified prophylactic measure
Z79.899	Other long term (current) drug therapy [prophylactic]

Anti-hemophilic factor (factor VIII) Recombinant: Advate, Afstyla, Kogenate FS, Kovaltry, Novoeight, Nuwiq, Recombinate, Xyntha/Xyntha Solofuse

HCPCS	Description
J7192	Factor VIII (Anti-hemophilic factor, recombinant) per IU, not otherwise specified [Advate, Kogenate-FS, Recombinate]
J7182	Injection, factor VIII, (Anti-hemophilic factor, recombinant), (Novoeight), per IU
J7185	Injection, factor VIII (Anti-hemophilic factor, recombinant) (Xyntha) (Xyntha Solofuse), per IU
J7209	Injection, factor VIII, (Anti-hemophilic factor, recombinant), (Nuwiq), 1 I.U.
J7210	Injection, factor VIII, (Anti-hemophilic factor, recombinant), (Afstyla), 1 I.U.
J7211	Injection, factor VIII, (Anti-hemophilic factor, recombinant), (Kovaltry), 1 I.U

ICD-10	Description
D66	Hereditary factor VIII deficiency [hemophilia A]
D68.00-D68.09	Von Willebrand's disease
D68.311	Acquired hemophilia
D68.318	Other hemorrhagic disorder due to intrinsic circulating anticoagulants, antibodies, or inhibitors
D68.4	Acquired coagulation factor deficiency
Z29.8	Encounter for other specified prophylactic measure
Z79.899	Other long term (current) drug therapy [prophylactic]

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Anti-hemophilic factor (factor VIII) – Long acting: Recombinant, pegylated (Adynovate); Jivi (damoctocog alfa pego); Recombinant Anti-hemophilic Factor, Fc Fusion Protein (Elocatate), Factor VIII Recombinant, glycopegylated (Esperoct), glycopegylated (Esperoct), Factor VIII Recombinant Antihemophilic Factor, Fc-VWF-XTEN Fusion Protein (Altuviiio)

HCPCS	Description
J7204	Injection, factor viii, (antihemophilic factor (recombinant), glycopegylated-exei, per iu [Esperoct]
J7205	Injection, factor VIII Fc fusion protein, (recombinant), per IU [Eloctate]
J7207	Injection, factor VIII, (anti-hemophilic factor, recombinant), pegylated, 1 I.U. [Adynovate]
J7208	Injection, factor viii, (antihemophilic factor, recombinant), pegylated-aucl, [Jivi], 1 i.u.
J7214	Injection, Factor VIII/von Willebrand factor complex, recombinant (Altuviiio), per Factor VIII IU

ICD-10	Description
D66	Hereditary factor VIII deficiency [hemophilia A]
D68.00-D68.09	Von Willebrand's disease
D68.311	Acquired hemophilia
Z29.8	Encounter for other specified prophylactic measure
Z79.899	Other long term (current) drug therapy [prophylactic]

Anti-hemophilic Factor VIII Recombinant, Porcine Sequence (Obizur)

HCPCS	Description
J7188	Injection, factor VIII (Anti-hemophilic factor, recombinant), (Obizur), per I.U.
J7191	Factor VIII, Anti-hemophilic factor (porcine), per IU

ICD-10	Description
D68.318	Other hemorrhagic disorder due to intrinsic circulating anticoagulants, antibodies, or inhibitors
D68.4	Acquired coagulation factor deficiency
D68.311	Acquired hemophilia

Anti-hemophilic Factor VIII/Von Willebrand Factor Complex (Alphanate, Humate-P, Wilate)

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HCPCS	Description
J7183	Injection, Von Willebrand factor complex (human) 1 IU VWF:RCO [Wilate]
J7186	Injection, antihemophilic factor VIII/Von Willebrand factor complex (human), per factor VIII I.U. [Alphanate]
J7187	Injection, Von Willebrand factor complex, per IU, VWF:RCO [Humate-P]

ICD-10	Description
D66	Hereditary factor VIII deficiency [hemophilia A]
D68.00-D68.09	Von Willebrand's disease
D68.311	Acquired hemophilia
D68.318	Other hemorrhagic disorder due to intrinsic circulating anticoagulants, antibodies, or inhibitors
D68.4	Acquired coagulation factor deficiency
Z29.8	Encounter for other specified prophylactic measure
Z79.899	Other long term (current) drug therapy [prophylactic]

Von Willebrand factor, Recombinant (Vonvendi)

HCPCS	Description
J7179	Injection, Von Willebrand factor (recombinant), (Vonvendi), 1 I.U. VWF:RCO

ICD-10	Description
D68.00-D68.09	Von Willebrand's disease
D68.311	Acquired hemophilia
Z29.8	Encounter for other specified prophylactic measure

Hemlibra (emicizumab) - Anti-hemophilic bispecific factor --- Factor IXa and Factor X

HCPCS	Description
J7170	Injection, emicizumab-kxwh, 0.5 mg [Hemlibra] [Note: code effective 01/01/2019]

ICD-10	Description
D66	Hereditary factor VIII deficiency [hemophilia A]
D68.00-D68.09	Von Willebrand's disease
D68.311	Acquired hemophilia
Z29.8	Encounter for other specified prophylactic measure
Z79.899	Other long term (current) drug therapy [prophylactic]

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Federal and state laws or requirements, contract language, and Plan utilization management programs or polices may take precedence over the application of this clinical criteria.

Medical Policy

Healthcare Services Department

Policy Name	Policy Number	Scope
Agents for Hemophilia A and Von Willebrand Disease	MP-RX-FP-02-23	☒ MMM MA ☒ MMM Multihealth

Policy History

Revision Type	Summary of Changes	P&T Approval Date	MPCC Approval Date
Policy Inception	Elevance Health's Medical Policy adoption.	N/A	11/30/2023

Revised: 10/01/2023