Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

Antihemophilic and Clotting Factors

Override(s)	Approval Duration
Prior Authorization	1 year

Medications	Quantity Limit
ADVATE	N/A
Adynovate	N/A
Afstyla	N/A
Alphanate	N/A
AlphaNine SD	N/A
Alprolix	N/A
BEBULIN	N/A
BeneFix	N/A
Coagadex	N/A
Corifact	N/A
Eloctate	N/A
Esperoct	N/A
FEIBA	N/A
Fibryga	N/A
Helixate FS	N/A
Hemlibra	N/A
HEMOFIL M	N/A
HUMATE-P	N/A
Idelvion	N/A
Ixinity	N/A
Jivi	N/A
Koate	N/A
Koate-DVI	N/A
Kogenate FS	N/A

This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

Kovaltry	N/A
Monoclate-P	N/A
Mononine	N/A
Novoeight	N/A
NovoSeven RT	N/A
Nuwiq	N/A
Obizur	N/A
Profilnine SD	N/A
Rebinyn	N/A
RECOMBINATE	N/A
RiaSTAP	N/A
RIXUBIS	N/A
TRETTEN	N/A
Vonvendi	N/A
Wilate	N/A
Xyntha	N/A
Xyntha Solufuse	N/A

APPROVAL CRITERIA

FEIBA (Anti-inhibitor Coagulant Complex)

Requests for FEIBA may be approved if the following criteria are met:

I. Individual has a diagnosis of hemophilia A or B with inhibitors to Factor VIII or Factor IX; **AND**

II. Individual is using for one of the following:
   A. Treatment of bleeding episodes; **OR**
   B. Peri-procedural operative management for surgical, invasive or interventional radiology procedures; **OR**
   C. Routine prophylaxis to prevent or reduce the frequency of bleeding episodes.

Feiba (Anti-inhibitor Coagulant Complex) may **not** be approved for the following:

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

I. Individual is using to treat bleeding episodes resulting from coagulation factor deficiencies in the absence of inhibitors to coagulation Factor VIII or coagulation Factor IX; **OR**
II. When the above criteria are not met and for all other indications.

Note: FEIBA (anti-inhibitor coagulant complex) has a black box warning for thrombotic and thromboembolic events, which have been reported during post marketing surveillance following infusion, particularly following the administration of high doses and/or in individuals with thrombotic risk factors.

NovoSeven RT (Factor VIIa Recombinant)

Requests for NovoSeven RT (Factor VIIa recombinant) may be approved if the following criteria are met:

I. Individual has one of the following diagnoses:
   A. Hemophilia A or B with inhibitors to Factor VIII or Factor IX; **OR**
   B. Acquired hemophilia; **OR**
   C. Congenital Factor VII deficiency;

AND

II. Individual is using for one of the following:
   A. Treatment of bleeding episodes; **OR**
   B. Prevention of bleeding in surgical interventions or invasive procedures.

OR

III. Individual has a diagnosis of Glansmann's thrombasthenia; **AND**
IV. Individual is using for the treatment of bleeding episodes and peri-operative management related to diagnosis; **AND**
V. Individual has documented refractoriness to platelet transfusions with or without antibodies to platelets;

Requests for NovoSeven RT (Factor VIIa recombinant) may not be approved when the above criteria are not met and for all other indications.

Note: NovoSeven, NovoSeven RT [coagulation Factor VIIa (recombinant)] has a black box warning for serious arterial and venous thrombotic events following administration. Individuals should be monitored for signs and symptoms of activation of the coagulation system and for thrombosis.

Hemofil M, Koate-DVI, Monoclate P (Factor VIII Human plasma-derived)

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

Requests for Hemofil M, Koate-DVI, or Monoclate-P (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 2016 update, Srivastava 2013);

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) (US National Hemophilia Foundation 2014, Srivastava 2013); **AND**
IX. Individual has documented history of one of the following (US National Hemophilia Foundation 2014, Srivastava 2013):
    A. One or more episodes of spontaneous bleeding into joint; **OR**
    B. One or more episodes of spontaneous bleeding into the central nervous system; **OR**
    C. Four or more episodes of soft tissue bleeding in an 8 week period.

Requests for Koate-DVI or Monoclate-P (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.

Hemofil M, Koate-DVI, or Monoclate-P (Factor VIII, human plasma-derived) may not be approved for the following:

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

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.

Advate, Afstyla, Helixate FS, Kogenate FS, Kovaltry, Novoeight, Nuwiq, Recombinate, Xyntha (Factor VIII Recombinant)

Requests for Advate, Afstyla, Helixate FS, Kogenate FS, Kovaltry, Novoeight, Nuwiq, Recombinate, or Xyntha (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 (VWD); **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 (VWD); **AND**
VIII. Individual is using for peri-procedural management for surgical, invasive or interventional radiology procedures; **AND**
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).

Requests for Advate, Afstyla, Helixate FS, Kogenate FS, Kovaltry, Novoeight, Nuwiq, or Xyntha (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**

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

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) (US National Hemophilia Foundation 2014, Srivastava 2013);

OR

III. Individual has 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) (US National Hemophilia Foundation 2014, Srivastava 2013); **AND**
IV. Individual is using as routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
V. Individual has documented history of one of the following:
    A. One or more episodes of spontaneous bleeding into joint; **OR**
    B. One or more episodes of spontaneous bleeding into the central nervous system; **OR**
    C. Four or more episodes of soft tissue bleeding in an 8 week period.

Requests for Helixate FS or 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.

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).

Advate, Afstyla, Helixate FS, Kogenate FS, Kovaltry, Novoeight, Nuwiq, Recombinate, or Xyntha (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.

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

Long Acting Agents [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), or Esperoct (Factor VIII Recombinant, glycopegylated)]

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), or Esperoct (Factor VIII Recombinant, glycopegylated) 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 2016 update, Srivastava 2013); **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) (US National Hemophilia Foundation 2014, Srivastava 2013); **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 there is documented history of one of the following:
        1. Individual has had one or more episodes of spontaneous bleeding into joint; **OR**
        2. Individual has had one or more episodes of spontaneous bleeding into the central nervous system; **OR**
        3. Individual has had four or more episodes of soft tissue bleeding in an 8 week period;

AND

VI. If using Jivi, individual is 12 years of age or older and has been previously treated with factor VIII.

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

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), or Esperoct (Factor VII Recombinant, glycopegylated) 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) - Anti-hemophilic bispecific factor - Factor IXa and Factor X Prior Authorization

Requests for Hemlibra (emicizumab) 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) (US National Hemophilia Foundation 2014, Srivastava 2013); **AND**
II. Individual is using for routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
III. Documentation of one of the following:
    A. Discontinuation of factor VIII agents being used for routine prophylaxis after the first week of Hemlibra initiation; **OR**
    B. Discontinuation of bypassing agents (i.e., NovoSeven RT, FEIBA) 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/dl) (US National Hemophilia Foundation 2014, Srivastava 2013); **AND**
V. Individual is using for routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
VI. Individual has documented history of one of the following:
    A. One or more episodes of spontaneous bleeding into joint; **OR**
    B. One or more episodes of spontaneous bleeding into the central nervous system; **OR**
    C. Four or more episodes of soft tissue bleeding in an 8 week period; **AND**
VII. Documentation of one of the following:
    A. Discontinuation of factor VIII agents being used for routine prophylaxis after the first week of Hemlibra initiation; **OR**

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

B. Discontinuation of bypassing agents (i.e., NovoSeven RT, FEIBA) being used for routine prophylaxis after 24 hours of Hemlibra initiation.

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

Obizur (Factor VIII Recombinant, Porcine Sequence)

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.

Requests for 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)

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 severe von Willebrand disease (VWD); **OR**
II. Individual has a diagnosis of mild to moderate VWD 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.

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:

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

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 2016 update, Srivastava 2013); **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) (US National Hemophilia Foundation 2014, Srivastava 2013); **AND**
VI. Individual is using for routine prophylaxis to prevent or reduce the frequency of bleeding episodes when there is documented history of one of the following:
    1. Individual has had one or more episodes of spontaneous bleeding into joint; **OR**
    2. Individual has had one or more episodes of spontaneous bleeding into the central nervous system; **OR**
    3. Individual has had four or more episodes of soft tissue bleeding in an 8 week period.

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 of acquired Factor VIII deficiency; **AND**
II. Individual is using for the treatment 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.

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

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.

Requests for 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)

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 (VWD); **OR**
   B. Individual has a diagnosis of mild to moderate VWD and use of desmopressin is known or suspected to be inadequate;

AND

III. Individual is using to treat spontaneous or trauma-induced bleeding episodes, or for peri-procedural management for surgical, invasive or interventional radiology procedures.

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

Alphanine SD or Mononine (Human plasma-derived, Coagulation Factor IX)

Requests for Alphanine SD or Mononine (Human plasma derived, Coagulation Factor IX) may be approved if the following criteria are met:

I. Individual has a diagnosis of hemophilia B (also called factor IX deficiency or Christmas disease); **AND**
II. Individual is using for the treatment of bleeding episodes;

OR

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

III. Individual has a diagnosis of severe hemophilia B (defined as less than 1 IU/dL or 1% endogenous Factor IX) (NHF 2016 update, Srivastava 2013);
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 B (defined as endogenous Factor IX less than 40 IU/dL [less than 40%], but greater than or equal to 1 IU/dL) (US National Hemophilia Foundation 2014, Srivastava 2013); **AND**
VI. Individual is using for routine prophylaxis to prevent or reduce the frequency of bleeding episodes;
AND
A. Individual has documented history of one of the following:
1. One or more episodes of spontaneous bleeding into joint; **OR**
2. One or more episodes of spontaneous bleeding into the central nervous system; **OR**
3. Four or more episodes of soft tissue bleeding in an 8 week period.

Alphanine SD or Mononine (Human plasma derived, Coagulation Factor IX) may **not** be approved for the following:

I. Treatment or reversal of coumarin-induced anticoagulation; **OR**
II. Hemorrhagic state or coagulopathy associated with liver dysfunction; **OR**
III. Treatment of individuals with hemophilia A with inhibitors to factor VIII; **OR**
IV. Replacement therapy of other clotting factors which include factors II, VII, and X; **OR**
V. When the above criteria are not met and for all other indications.

Bebulin or Profilnine SD (Human plasma-derived, Factor IX Complex)

Requests for Bebulin or Profilnine SD (Human plasma-derived, Factor IX Complex) may be approved if the following criteria are met:

I. Individual has a diagnosis of hemophilia B (also called factor IX deficiency or Christmas disease); **AND**
II. Individual is using for the treatment of bleeding episodes;

OR
III. Individual has a diagnosis of severe hemophilia B (defined as less than 1 IU/dL or 1% endogenous Factor IX) (NHF 2016 update, Srivastava 2013); **AND**
IV. Individual is using as routine prophylaxis to prevent or reduce the frequency of bleeding episodes;

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

OR

V. Individual has a diagnosis of mild to moderate hemophilia B (defined as endogenous Factor IX less than 40 IU/dL [less than 40%], but greater than or equal to 1 IU/dL) (US National Hemophilia Foundation 2014, Srivastava 2013); **AND**

VI. Individual is using for routine prophylaxis to prevent or reduce the frequency of bleeding episodes;
AND

VII. Individual has documented history of one of the following:
   A. One or more episodes of spontaneous bleeding into joint; **OR**
   B. One or more episodes of spontaneous bleeding into the central nervous system; **OR**
   C. Four or more episodes of soft tissue bleeding in an 8 week period.

Bebulin or Profilnine SD (Human plasma-derived, Factor IX Complex) may **not** be approved for the following:

I. Individual has a diagnosis of Factor VII deficiency; **OR**
II. When the above criteria are not met and for all other indications.

Benefix, Ixinity, or Rixubis (Recombinant Factor IX)

Requests for Benefix or Rixubis (Recombinant Factor IX) may be approved if the following criteria are met:

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

OR

III. Individual has a diagnosis of severe hemophilia B (defined as less than 1 IU/dL or 1% endogenous Factor IX) (US National Hemophilia Foundation 2014, Srivastava 2013); **AND**
IV. Individual is using as routine prophylaxis to prevent or reduce the frequency of bleeding episodes;

OR

V. Individual has a diagnosis of mild to moderate hemophilia B (defined as endogenous Factor IX less than 40 IU/dL [less than 40%], but greater than or equal to 1 IU/dL) (US National Hemophilia Foundation 2014, Srivastava 2013); **AND**

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

VI. Individual is using for routine prophylaxis to prevent or reduce the frequency of bleeding episodes;
AND
VII. Individual has documented history of one of the following:
1. One or more episodes of spontaneous bleeding into joint; **OR**
2. One or more episodes of spontaneous bleeding into the central nervous system; **OR**
3. Four or more episodes of soft tissue bleeding in an 8 week period.

Requests for Ixinity (Recombinant Factor IX) may be approved if the following criteria are met:

I. Individual is 12 years of age or older; **AND**
II. Individual has a diagnosis of hemophilia B (also called factor IX deficiency or Christmas disease);
AND
III. Individual is using for one of the following:
A. The treatment of bleeding episodes; **OR**
B. Peri-procedural management for surgical, invasive or interventional radiology procedures;

OR
IV. Individual has a diagnosis of severe hemophilia B (defined as less than 1 IU/dL or 1% endogenous Factor IX) (NHF 2016 update, Srivastava 2013); **AND**
V. Individual is using as routine prophylaxis to prevent or reduce the frequency of bleeding episodes;

OR
VI. Individual has a diagnosis of mild to moderate hemophilia B (defined as endogenous Factor IX less than 40 IU/dL [less than 40%], but greater than or equal to 1 IU/dL) (NHF 2016 update, Srivastava 2013); **AND**
VII. Individual is using for routine prophylaxis to prevent or reduce the frequency of bleeding episodes; **AND**
VIII. Individual has documented history of one of the following:
A. One or more episodes of spontaneous bleeding into joint; **OR**
B. One or more episodes of spontaneous bleeding into the central nervous system; **OR**
C. Four or more episodes of soft tissue bleeding in an 8 week period.

Benefix, Ixinity, Rixubis (Recombinant Factor IX) may not be approved for the following:

I. Treatment of other factor deficiencies (for example factors II, VII, VIII and X); **OR**
II. Treatment of individuals with hemophilia A with inhibitors to factor VIII; **OR**
III. To reverse coumarin-induced anticoagulation; **OR**

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

IV. Treatment of bleeding due to low levels of liver-dependent coagulation factors.

Ixinity, Rixubis (Recombinant Factor IX) may not be approved for the following:

I. Using for the induction of immune tolerance in individuals with hemophilia B.

Requests for Benefix, Ixinity, Rixubis (Recombinant Factor IX) may not be approved when the above criteria are not met and for all other indications.

Idelvion (Recombinant Long-Acting, Albumin Fusion Protein Coagulation Factor IX), Alprolix (Recombinant, Fc Fusion Protein Coagulation Factor IX), or Rebinyn (Recombinant, glycoPEGylated Coagulation Factor IX)

Requests for Idelvion (Recombinant Long-Acting, Albumin Fusion Protein Coagulation Factor IX), Alprolix (Recombinant, Fc Fusion Protein Coagulation Factor IX), or Rebinyn (Recombinant, glycoPEGylated Coagulation Factor IX) may be approved if the following criteria are met:

I. Individual has a diagnosis of severe hemophilia B (also called factor IX deficiency or Christmas disease); **AND**
II. Individual has less than 1 IU/dL (less than 1%) endogenous Factor IX (NHF 2016 update, Srivastava 2013); **AND**
III. Individual is using for one of the following:
    A. The treatment of 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 (excluding Rebinyn [Recombinant GlycoPEGylated Coagulation Factor IX]);

OR

IV. Individual has a diagnosis of mild to moderate hemophilia B; **AND**
V. Individual has endogenous Factor IX level less than 40 IU/dL (less than 40%) but greater than or equal to 1 IU/dL (US National Hemophilia Foundation 2014, Srivastava 2013); **AND**
VI. Individual is using for one of the following:
    A. Individual is using for the treatment of bleeding episodes; **OR**
    B. Individual is using for peri-procedural management for surgical, invasive or interventional radiology procedures; **OR**
    C. Individual is using for routine prophylaxis to prevent or reduce the frequency of bleeding episodes when there is documented history of one of the following (excluding Rebinyn [Recombinant GlycoPEGylated Coagulation Factor IX]):
        1. Individual has had one or more episodes of spontaneous bleeding into

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

joint; **OR**

2. Individual has had one or more episodes of spontaneous bleeding into the central nervous system; **OR**
3. Individual has had four or more episodes of soft tissue bleeding in an 8 week period.

Idelvion (Recombinant Long-Acting, Albumin Fusion Protein Coagulation Factor IX), Alprolix (Recombinant, Fc Fusion Protein Coagulation Factor IX), or Rebinyn (Recombinant, GlycoPEGylated Coagulation Factor IX) may not be approved for the following:

I. Using for the induction of immune tolerance in individuals with hemophilia B.

Rebinyn (Recombinant, GlycoPEGylated Coagulation Factor IX) may **not** be approved for the following:

I. Individual is using for prophylaxis in the prevention or reduction of the frequency of bleeding episodes with hemophilia B.

Requests for Idelvion (Recombinant Long-Acting, Albumin Fusion Protein Coagulation Factor IX), Alprolix (Recombinant, Fc Fusion Protein Coagulation Factor IX), or Rebinyn (Recombinant, glycoPEGylated Coagulation Factor IX) may not be approved when the above criteria are not met and for all other indications.

Coagadex (Human-plasma derived Coagulation Factor X)

Requests for Coagadex (Human-plasma derived Coagulation Factor X) may be approved if the following criteria are met:

I. Individual has a diagnosis of severe or moderate hereditary Factor X deficiency (defined as less than 5 IU/dL or 5% endogenous Factor X) (US National Hemophilia Foundation 2014, Srivastava 2013); **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 (excluding perioperative management of bleeding in major surgery in individuals with severe hereditary Factor X deficiency); **OR**
    C. Routine prophylaxis to prevent or reduce the frequency of bleeding episodes;

OR

III. Individual has a diagnosis of mild hereditary Factor X deficiency (defined as greater than or equal to 5 IU/dL or 5% endogenous Factor X) (US National Hemophilia Foundation 2014, Srivastava 2013); **AND**

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

IV. 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 there is documented history of one of the following:
      1. One or more episodes of spontaneous bleeding into joint; **OR**
      2. One or more episodes of spontaneous bleeding into the central nervous system; **OR**
      3. Four or more episodes of soft tissue bleeding in an 8 week period.

Coagadex (Human-plasma derived Coagulation Factor X) may **not** be approved for the following:

I. Individual with severe hereditary Factor X deficiency is using for perioperative management of bleeding in major surgery; **OR**
II. When the above criteria are not met and for all other indications.

Tretten or Corifact (Factor XIII)

Requests for Corifact (Human Plasma-derived, Factor XIII) may be approved if the following criteria are met:

I. Individual has a diagnosis of Factor XIII deficiency;
AND
II. Individual is using for routine prophylactic treatment to prevent or reduce the frequency of bleeding episodes; **OR**
III. Individual is using for peri-procedural management for surgical, invasive or interventional radiology procedures.

Requests for Tretten (Recombinant Factor XIII A-Subunit) may be approved if the following criteria are met:

I. Individual has a diagnosis of congenital Factor XIII A-Subunit deficiency; **AND**
II. Individual is using as routine prophylaxis for bleeding.

Corifact (Human Plasma-derived, Factor XIII) or Tretten (Recombinant Factor XIII A-Subunit) may **not** be approved for the following:

I. Individual with congenital Factor XIII B-subunit deficiency; **OR**

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

II. When the above criteria are not met and for all other indications.

RiaSTAP (Human plasma-derived Fibrinogen concentrate) or Fibryga (Human fibrinogen)

Requests for RiaSTAP (Human plasma-derived Fibrinogen concentrate) or Fibryga (Human fibrinogen) may be approved if the following criteria are met:

I. Individual has a diagnosis of congenital fibrinogen deficiency (afibrinogenemia or hypofirbinogenemia); **AND**
II. Individual is using for the treatment of acute bleeding episodes.

RiaSTAP (Human plasma-derived Fibrinogen concentrate) or Fibryga (Human fibrinogen) may **not** be approved for the following:

I. Individual has a diagnosis of dysfibrinogenemia; **OR**
II. When the above criteria are not met and for all other indications.

State Specific Mandates		
State Name N/A	Date Effective N/A	Mandate details (including specific bill if applicable) N/A

Key References:

1. Centers for Disease Control and Prevention. Hemophilia Facts. Updated September 6 ,2018. Available at: http://www.cdc.gov/ncbddd/hemophilia/facts.html. Accessed on September 18, 2019.
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3. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: September 19, 2019.
4. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
5. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2019; Updated periodically.
6. National Hemophilia Foundation (NHF). Available at: http://www.hemophilia.org/. Accessed on September 18, 2019.
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8. Srivastava A, Brewer AK, Mauser-Bunschoten EP, et al.; Treatment Guidelines Working Group on Behalf of the World Federation Of Hemophilia. Guidelines for the management of hemophilia. Haemophilia. 2013; 19(1):e1-e47.

This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.

Market Applicability							
Market	**DC**	**GA**	**KY**	**MD**	**NJ**	**NY**	**WA**
Applicable	X	X	X	X	X	X	NA

9. Vonvendi – von Willebrand factor (recombinant) Package Insert. Baxalta US Inc. 2018. Available at file:///C:/Users/AF38863/Downloads/20190215_53b6d198-6175-4a26-8606-dc0d2a0f12d6%20(1).pdf Accessed on September 18, 2019.

This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.