

Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

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Antihemophilic and Clotting Factors

CG-DRUG-78.

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
Andexxa	N/A
BEBULIN	N/A
BeneFix	N/A
Coagadex	N/A
Corifact	N/A
Eloctate	N/A
FEIBA	N/A
Fibryna	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

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	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

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Koate-DVI	N/A
Kogenate FS	N/A
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

Anti-inhibitor Coagulant Complex (FEIBA)

Requests for anti-inhibitor coagulant complex agents (FEIBA) **may be approved** to treat individuals with hemophilia A or B with inhibitors to Factor VIII or Factor IX when the following criteria are met:

- I. Treatment of bleeding episodes; **or**

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Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

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- II. Peri-procedural operative management for surgical, invasive or interventional radiology procedures; **or**
- III. Routine prophylaxis to prevent or reduce the frequency of bleeding episodes.

Anti-inhibitor coagulant complex **may not be approved** when the above criteria are not met and for all other indications including, but not limited to treatment of bleeding episodes resulting from coagulation factor deficiencies in the absence of inhibitors to coagulation Factor VIII or coagulation Factor IX.

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.

Factor VIIa Recombinant (NovoSeven RT)

Requests for Factor VIIa recombinant coagulation agents (NovoSeven RT) **may be approved** for the following:

- I. For treatment of bleeding episodes when the following criteria are met:
 - a. Individual has hemophilia A or B with inhibitors to Factor VIII or Factor IX; **or**
 - b. Individual has acquired hemophilia; **or**
 - c. Individual has congenital Factor VII deficiency.

OR

- II. In the prevention of bleeding in surgical interventions or invasive procedures for the following:
 - a. Individual has hemophilia A or B with inhibitors to Factor VIII or Factor IX; **or**
 - b. Individual has acquired hemophilia; **or**
 - c. Individual has congenital Factor VII deficiency.

OR

- III. For the treatment of bleeding episodes and peri-operative management in individuals with Glanzmann’s thrombasthenia and a documented refractoriness to platelet transfusions with or without antibodies to platelets.

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

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	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

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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.

Antihemophilic factor (factor VIII) Human plasma-derived (HEMOFIL M, Koate-DVI, Monoclate-P)

Requests for antihemophilic factor (Factor VIII) human plasma-derived agents (HEMOFIL M, Koate-DVI, Monoclate-P) **may be approved** for the following:

- I. For the treatment of bleeding episodes in an individual with hemophilia A and factor VIII deficiency. **OR**
- II. As routine prophylaxis to prevent or reduce the frequency of bleeding episodes when the following criteria are met:
 - a. Individual has severe hemophilia A (defined as less than 1 International Unit per deciliter [IU/dL] or 1% endogenous Factor VIII); **or**
 - b. 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); **and**
 - c. When the individual has documented history of one of the following:
 - i. 1 or more episodes of spontaneous bleeding into joint; **or**
 - ii. 1 or more episodes of spontaneous bleeding into the central nervous system; **or**
 - iii. 4 or more episodes of soft tissue bleeding in an 8 week period.

Requests for antihemophilic factor (Factor VIII) human plasma-derived agents (Koate-DVI, Monoclate-P) **may be approved** for the following:

- I. As peri-procedural management for surgical, invasive or interventional radiology procedures in an individual with hemophilia A and factor VIII deficiency.

Antihemophilic factor (Factor VIII) human plasma-derived agents (HEMOFIL M, Koate-DVI, Monoclate-P) **may not be approved** when the above criteria are not met including, but not limited to treatment of individuals with von Willebrand disease (VWD).

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	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

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Antihemophilic factor (factor VIII) Recombinant (ADVATE, Afstyla, Helixate FS, Kogenate FS, Kovaltry, Novoeight, Nuwiq, RECOMBINATE, Xyntha)

Requests for Antihemophilic factor (Factor VIII) recombinant agents (ADVATE, Afstyla, Helixate FS, Kogenate FS, Kovaltry, Novoeight, Nuwiq, RECOMBINATE, Xyntha) **may be approved** for the following:

- I. For the treatment of bleeding episodes in an individual with hemophilia A and factor VIII deficiency; **OR**
- II. For the treatment of bleeding episodes in an individual with von Willebrand disease (VWD) when the following criteria are met:
 - a. Antihemophilic Factor VIII Recombinant is used in combination with recombinant von Willebrand factor, when medically necessary as per Vonvendi criteria below; **AND**
 - b. Baseline factor VIII levels are less than 40 IU/dL [less than 40%] or are unknown.
- III. As peri-procedural management for surgical, invasive or interventional radiology procedures for an individual with hemophilia A and Factor VIII deficiency.

Requests for Antihemophilic Factor VIII Recombinant (*ADVATE, Afstyla, Helixate FS, Kovaltry, Novoeight, Nuwiq, Xyntha*) **may be approved** as routine prophylaxis to prevent or reduce the frequency of bleeding episodes when the following criteria are met (I alone **OR** II&III):

- I. Individual has severe hemophilia A (defined as less than 1 International Unit per deciliter [IU/dL] or 1% endogenous Factor VIII);

OR

- II. 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); **and**
- III. When the individual has documented history of one of the following:
 - a. 1 or more episodes of spontaneous bleeding into joint; **or**
 - b. 1 or more episodes of spontaneous bleeding into the central nervous system; **or**
 - c. 4 or more episodes of soft tissue bleeding in an 8 week period.

Requests for Antihemophilic Factor VIII Recombinant (*Helixate FS, Kogenate FS*) **may be approved** for the following:

- I. As *routine prophylaxis* for children (age 0-16 years) with hemophilia A and factor VIII deficiency to reduce the risk of joint damage in those without pre-existing joint damage.

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	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

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Requests for Antihemophilic Factor VIII Recombinant (*Recombinate*) **may be approved** for the following:

- I. As treatment of individuals with acquired Factor VIII inhibitors not exceeding 10 Bethesda Unit (BU) per milliliter (mL).

Antihemophilic Factor VIII Recombinate (*ADAVATE, Helixate FS, Kogenate, Kovaltry, Novoeight, Nuwiq, Recombinate, Xyntha*) **may not be approved** when the above criteria are not met and for all other indications.

Antihemophilic Factor (factor VIII) – Long-Acting Recombinant, pegylated (Adynovate); Antihemophilic factor (recombinant), PEGylated-aucl (Jivi)*; Recombinant Antihemophilic Factor Fc Fusion Protein (Eloctate)

Requests for Antihemophilic Factor (factor VIII) – Long-Acting Recombinant, pegylated (Adynovate); Antihemophilic factor (recombinant), PEGylated-aucl (Jivi); Recombinant Antihemophilic Factor Fc Fusion Protein (Eloctate) **may be approved** for the following:

- I. Individuals with *severe* hemophilia A (congenital factor VIII deficiency); **AND**
 - a. Individual has less than 1 International Unit per deciliter (IU/dL) (less than 1%) endogenous factor VIII; **and**
 - b. Use is planned for one of the following indications:
 - i. Control and prevention of acute bleeding episodes; **or**
 - ii. Peri-procedural management for surgical, invasive or interventional radiology procedures; **or**
 - iii. Routine prophylaxis to prevent or reduce the frequency of bleeding episodes.

OR

- II. Individuals with *mild to moderate* hemophilia A (congenital factor VIII deficiency); **AND**
 - a. Individual has endogenous factor VIII level less than 40 IU/dl (less than 40%) but greater than or equal to 1 IU/dl; **and**
 - b. Use is planned for one of the following indications:
 - i. Control of acute bleeding episodes; **or**
 - ii. Peri-procedural management for surgical, invasive or interventional radiology procedures; **or**

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Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

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- iii. Routine prophylaxis to prevent or reduce the frequency of bleeding episodes when the member has documented history of one of the following:
 - 1) 1 or more episodes of spontaneous bleeding into joint; **or**
 - 2) 1 or more episodes of spontaneous bleeding into the central nervous system; **or**
 - 3) 4 or more episodes of soft tissue bleeding in an 8 week period.

***Requests for Jivi may not be approved for patients under 12 years of age.**

***Jivi is not indicated for use in previously untreated patients.**

Requests for Antihemophilic Factor (factor VIII) Recombinant, pegylated (Adynovate); Antihemophilic factor (recombinant), PEGylated-aucl (Jivi); Recombinant Antihemophilic Factor Fc Fusion Protein (Eloctate) **may not be approved** when the above criteria are not met and for all other indications including, but not limited to treatment of individuals with von Willebrand Disease

Antihemophilic bispecific factor (Factor IXa- and Factor X-), Emicizumab (Hemlibra)

Requests for Antihemophilic bispecific factor (Factor IXa- and Factor X-), Emicizumab (Hemlibra) **may be approved** for the following:

- I. Emicizumab (Hemlibra) is considered medically necessary for individuals with severe hemophilia A (congenital factor VIII deficiency) when all of the following criteria are met:
 - a. Individual has less than 1 International Unit per deciliter (IU/dL) (less than 1%) endogenous factor VIII; **AND**
 - b. Individual has a documented history of a high-titer of factor VIII inhibitor (that is: greater than or equal to [\geq] 5 bethesda units [BU]) requiring treatment with episodic or prophylactic bypassing agents; **AND**
 - c. Use is planned for routine prophylaxis to prevent or reduce the frequency of bleeding episodes.
- II. Emicizumab (Hemlibra) is considered medically necessary for individuals with mild to moderate hemophilia A (congenital factor VIII deficiency) when all of the following criteria are met:
 - a. Individual has endogenous factor VIII level less than 40 IU/dl (less than 40%) but greater than or equal to 1 IU/dl; **AND**
 - b. Individual has a documented history of a high-titer of factor VIII inhibitor (that is: greater than or equal to [\geq] 5 bethesda units [BU]) requiring treatment with episodic or prophylactic bypassing agents; **AND**
 - c. Use is planned for routine prophylaxis to prevent or reduce the frequency of

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Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

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bleeding episodes when the member has documented history of one of the following:

- i. 1 or more episodes of spontaneous bleeding into joint; **OR**
- ii. 1 or more episodes of spontaneous bleeding into the central nervous system; **OR**
- iii. 4 or more episodes of soft tissue bleeding in an 8 week period.

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

Antihemophilic Factor (Recombinant), Porcine Sequence (Obizur)

Requests for Antihemophilic Factor (Recombinant), Porcine Sequence **may be approved** for the following:

- I. For treatment of bleeding episodes in adults with acquired hemophilia A.

Antihemophilic Factor (Recombinant), Porcine Sequence (Obizur) **may not be approved** when the above criteria are not met and for all other indications including, but not limited to:

- I. Treatment of individuals with congenital hemophilia A with Factor VIII deficiency
- II. Treatment of individuals with von Willebrand disease
- III. Treatment of individuals with acquired hemophilia A and baseline anti-porcine Factor VIII inhibitor titer greater than 20 BU/mL.

Antihemophilic Factor VIII/von Willebrand Factor Complex (Alphanate, Humate-P, Wilate)

Requests for Antihemophilic Factor VIII/von Willebrand Factor Complex (Alphanate, Humate-P, Wilate) **may be approved** as treatment for individuals with von Willebrand disease when the following criteria are met (I alone **OR** II & III):

- I. VWD is severe;

OR

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Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

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- II. VWD is mild to moderate and use of desmopressin is known or suspected to be inadequate; **and**
- III. Individual is being treated for either:
 - a. Spontaneous or trauma-induced bleeding episodes; **or**
 - b. Peri-procedural management for surgical, invasive or interventional radiology procedures.

Requests for Antihemophilic Factor VIII/von Willebrand Factor Complex (Alphanate, Humate-P) **may be approved** for the following:

- I. For treatment of bleeding episodes in an individual with hemophilia A and Factor VIII deficiency; **OR**
- II. As routine prophylaxis to prevent or reduce the frequency of bleeding episodes when the following criteria are met:
 - a. Individual has severe hemophilia A (defined as less than 1 International Unit per deciliter [IU/dL] or 1% endogenous Factor VIII); **OR**
 - b. 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); **AND**
 - c. When the individual has documented history of one of the following:
 - i. 1 or more episodes of spontaneous bleeding into joint; **OR**
 - ii. 1 or more episodes of spontaneous bleeding into the central nervous system; **OR**
 - iii. 4 or more episodes of soft tissue bleeding in an 8 week period.

Requests for Antihemophilic Factor VIII/von Willebrand Factor Complex (Alphanate) **may be approved** for the following:

- I. For treatment of bleeding episodes in an individual with acquired Factor VIII deficiency.

Antihemophilic Factor/von Willebrand Factor Complex (Alphanate, Humate-P, Wilate) **may not be approved** when the above criteria are not met and for any of the following:

- I. All other indications including, but not limited to prophylaxis therapy in individuals with VWD.
- II. Alphanate for individuals with severe VWD (Type 3) undergoing major surgery.
- III. Wilate for individuals with hemophilia A.

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Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

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von Willebrand factor (Recombinant) (Vonvendi)

Requests for von Willebrand factor (Recombinant) (Vonvendi) **may be approved** for the following:

- I. von Willebrand factor (Recombinant) is considered **medically necessary** as a treatment for adults (18 years of age and older) with von Willebrand disease when the following criteria are met:
 - a. VWD is *severe*; **or**
 - b. VWD is *mild to moderate* and use of desmopressin is known or suspected to be inadequate; **and**
 - c. Individual is being treated for spontaneous or trauma-induced bleeding episodes or peri-procedural management for surgical, invasive or interventional radiology procedures.

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

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

Requests for Human plasma-derived coagulation Factor IX (Alphanine SD, Mononine) **may be approved** for the following:

- I. For treatment of bleeding episodes in an individual with hemophilia B and Factor IX deficiency; **OR**
- II. As routine prophylaxis to prevent or reduce the frequency of bleeding episodes when the following criteria are met:
 - a. Individual has severe hemophilia B (defined as less than 1 IU/dL or 1% endogenous Factor IX); **or**
 - b. Individual has 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); **and**
 - c. When the member has documented history of one of the following:
 - i. 1 or more episodes of spontaneous bleeding into joint; **or**
 - ii. 1 or more episodes of spontaneous bleeding into the central nervous system; **or**
 - iii. 4 or more episodes of soft tissue bleeding in an 8 week period.

Human plasma-derived coagulation Factor IX (Alphanine SD, Mononine) **may not be approved** when the above criteria are not met, including but not limited to the following:

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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.

CRX-ALL-0292-18

Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

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- I. Treatment or reversal of coumarin-induced anticoagulation
- II. Hemorrhagic state or coagulopathy associated with liver dysfunction
- III. Treatment of individuals with hemophilia A with inhibitors to factor VIII
- IV. Replacement therapy of other clotting factors which include factors II, VII and X.

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

Requests for Human plasma-derived Factor IX complex (*Bebulin, Profilnine SD*) **may be approved** for the following:

- I. For treatment of bleeding episodes in an individual with hemophilia B (congenital factor IX deficiency or Christmas disease); **OR**
- II. As routine prophylaxis to prevent or reduce the frequency of bleeding episodes when the following criteria are met:
 - a. Individual has severe hemophilia B (defined as less than 1 IU/dL or 1% endogenous Factor IX); **or**
 - b. Individual has 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); **and**
 - c. When the member has documented history of one of the following:
 - i. 1 or more episodes of spontaneous bleeding into joint; **or**
 - ii. 1 or more episodes of spontaneous bleeding into the central nervous system; **or**
 - iii. 4 or more episodes of soft tissue bleeding in an 8 week period.

Human plasma-derived Factor IX complex (*Bebulin, Profilnine SD*) **may not be approved** when the above criteria are not met and for all other indications including, but not limited to use for treatment of individuals with Factor VII deficiency.

Factor IX Recombinant (*Benefix, Ixinity, RIXUBIS*)

Requests for Recombinant coagulation Factor IX (*Benefix, RIXUBUS*) **may be approved** to treat individuals with hemophilia B (congenital factor IX deficiency or Christmas disease) when the following criteria are met:

- I. To treat bleeding episodes; **or**
- II. For peri-procedural management for surgical, invasive or interventional radiology procedures.

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Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

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Requests for Recombinant coagulation Factor IX (Benefix, RIXUBIS) **may be approved** as routine prophylaxis to prevent or reduce the frequency of bleeding episodes when the following criteria are met (I alone **OR** II & III):

- I. Individual has severe hemophilia B (defined as less than 1 IU/dL or 1% endogenous Factor IX);

OR

- II. Individual has 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); **and**
- III. When the member has documented history of one of the following:
 - a. 1 or more episodes of spontaneous bleeding into joint; **or**
 - b. 1 or more episodes of spontaneous bleeding into the central nervous system; **or**
 - c. 4 or more episodes of soft tissue bleeding in an 8 week period.

Recombinant coagulation Factor IX (Ixinity) **may be approved** to treat individuals aged 12 years and older with hemophilia B (congenital factor IX deficiency or Christmas disease) when the following criteria are met:

- I. To treat bleeding episodes; **or**
- II. Peri-procedural management for surgical, invasive or interventional radiology procedures; **or**
- III. Routine prophylaxis to prevent or reduce the frequency of bleeding episodes when the following criteria are met:
 - a. Individual has severe hemophilia B (defined as less than 1 IU/dL or 1% endogenous Factor IX); **or**
 - b. Individual has 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); **and**
 - c. When the member has documented history of one of the following:
 - i. 1 or more episodes of spontaneous bleeding into joint; **or**
 - ii. 1 or more episodes of spontaneous bleeding into the central nervous system; **or**
 - iii. 4 or more episodes of soft tissue bleeding in an 8 week period.

Recombinant coagulation Factor IX (*Benefix, Ixinity, RIXUBIS*) **may not be approved** when the above criteria are not met and for all other indications including, but not limited to the following:

- I. Treatment of other factor deficiencies (for example factors II, VII, VIII and X)
- II. Treatment of individuals with hemophilia A with inhibitors to factor VIII

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Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

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- III. To reverse coumarin-induced anticoagulation
- IV. Treatment of bleeding due to low levels of liver-dependent coagulation factors.
- V. For the induction of immune tolerance in individuals with hemophilia B.

Coagulation Factor IX - Long-Acting Recombinant, Albumin Fusion Protein (*Idelvion*); Recombinant Coagulation Factor IX, Fc Fusion Protein (*Alprolix*); Recombinant Coagulation Factor IX, GlycoPEGylated (*Rebinyn*[®])

Requests for Coagulation Factor IX - Long Acting Recombinant, Albumin Fusion Protein (*Idelvion*); Recombinant Coagulation Factor IX, Fc Fusion Protein (*Alprolix*); Recombinant Coagulation Factor IX, GlycoPEGylated (*Rebinyn*) may be approved for the following:

- I. Individual has *severe* hemophilia B (congenital Factor IX deficiency); **AND**
 - a. Individual has less than 1 International Unit per deciliter (IU/dl) (less than 1%) endogenous factor IX; **and**
 - b. Use of rIX-FP is planned for one of the following indications:
 - i. Treatment of bleeding episodes; **or**
 - ii. Peri-procedural management for surgical, invasive or interventional radiology procedures; **or**
 - iii. Routine prophylaxis to prevent or reduce the frequency of bleeding episodes (excluding Recombinant Coagulation Factor IX, GlycoPEGylated [*Rebinyn*]).

OR

- II. Individuals has *mild to moderate* hemophilia B (congenital Factor IX deficiency); **AND**
 - a. Individual has endogenous factor IX level less than 40 International Units per deciliter (IU/dl) (less than 40%) but greater than or equal to 1 IU/dl; **and**
 - b. Use of rIX-FP is planned for one of the following indications:
 - i. Treatment of bleeding episodes; **or**
 - ii. Peri-procedural management for surgical, invasive or interventional radiology procedures; **or**
 - iii. Routine prophylaxis to prevent or reduce the frequency of bleeding episodes when the member has documented history of one of the following (excluding Recombinant Coagulation Factor IX, GlycoPEGylated [*Rebinyn*]):
 - 1) 1 or more episodes of spontaneous bleeding into joint; **or**
 - 2) 1 or more episodes of spontaneous bleeding into the central nervous system;
 - or**
 - 3) 4 or more episodes of soft tissue bleeding in an 8 week period.

Coagulation Factor IX - Long-Acting Recombinant, Albumin Fusion Protein (*Idelvion*); Recombinant Coagulation Factor IX, Fc Fusion Protein (*Alprolix*); Recombinant Coagulation

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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.

CRX-ALL-0292-18

Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

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Factor IX, GlycoPEGylated (*Rebinyn*) **may not be approved** when the above criteria are not met and for all other indications, including but not limited to induction of immune tolerance in individuals with hemophilia B.

Recombinant Coagulation Factor IX, GlycoPEGylated (*Rebinyn*) **may not be approved** for prophylactic use in the prevention or reduction of the frequency of bleeding episodes.

Coagulation Factor X, Human plasma-derived (Coagadex)

Requests for Human plasma derived coagulation Factor X (Coagadex) may be approved for individuals aged 12 years or older when the following criteria are met:

- I. Individual has severe or moderate hereditary Factor X deficiency (defined as less than 5 International Unit per deciliter (IU/dl) or 5% endogenous Factor X) **and** the factor is to be used for the treatment of bleeding episodes; **OR**
- II. Individual has mild hereditary Factor X deficiency (defined as greater than or equal to 5 International Unit per deciliter (IU/dl) or 5% endogenous Factor X) **and** the factor is to be used for peri-procedural management for surgical, invasive or interventional radiology procedures.

Human plasma derived coagulation Factor X (Coagadex) **may not be approved** when the above criteria are not met and for all other indications, including but not limited to perioperative management of bleeding in major surgery in individuals with moderate and severe hereditary Factor X deficiency.

Factor XIII (Corifact, TRETEN)

Requests for Human plasma-derived concentrate Factor XIII (Corifact) **may be approved** for individuals with Factor XIII deficiency for the following indications:

- I. As routine prophylactic treatment to prevent or reduce the frequency of bleeding episodes; **or**
- II. Peri-procedural management for surgical, invasive or interventional radiology procedures.

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	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

*FHK- Florida Healthy Kids

Requests for Recombinant coagulation Factor XIII A-Subunit (TRETEN) **may be** approved for the following:

- I. As routine prophylaxis for bleeding in individuals with congenital Factor XIII A-subunit deficiency.

Coagulation Factor XIII (Corifact, TRETEN) **may not be approved** when the above criteria are not met and for all other indications including, but not limited to treatment of individuals with congenital Factor XIII B-subunit deficiency.

Fibrinogen Concentrate, Human plasma-derived (RiaSTAP); Human fibrinogen (Fibryna)

Requests for Human plasma-derived fibrinogen concentrate (RiaSTAP) and human fibrinogen (Fibryna) **may be approved** for the following:

- I. For the treatment of acute bleeding episodes in individuals with congenital fibrinogen deficiency (that is, afibrinogenemia or hypofibrinogenemia).

Human plasma-derived fibrinogen concentrate (RiaSTAP) and human fibrinogen (Fibryna) **may not be approved** when the above criteria are not met and for all other indications including, but not limited to treatment of individuals with dysfibrinogenemia.

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

Key References:

1. Athale AH, Marcucci M, Iorio A. Immune tolerance induction for treating inhibitors in people with congenital haemophilia A or B. Cochrane Database Syst Rev. 2014;(4):CD010561.

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	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	NA

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2. Iorio A, Marchesini E, Marcucci M, et al. Clotting factor concentrates given to prevent bleeding and bleeding-related complications in people with hemophilia A or B. *Cochrane Database Syst Rev.* 2011;(9):CD003429.
3. Nichols WL, Hultin MB, James AH, et al. von Willebrand disease (VWD): evidence-based diagnosis and management guidelines, the National Heart, Lung, and Blood Institute (NHLBI) Expert Panel report (USA). *Haemophilia.* 2008; 14(2):171-232.
4. Simpson E, Lin Y, Stanworth S, et al. Recombinant factor VIIa for the prevention and treatment of bleeding in patients without haemophilia. *Cochrane Database Syst Rev.* 2012; (3):CD005011.
5. 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.
6. Yank V, Tuohy CV, Logan AC, et al. Comparative effectiveness of recombinant factor VIIa for off-label indications vs. usual care. Comparative effectiveness review No. 21. Rockville, MD: Agency for Healthcare Research and Quality. May 2010. Available at: https://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0047460/pdf/PubMedHealth_PMH0047460.pdf. Accessed on March 22, 2018.

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.