

Clinical Policy: Factor IX (Human, Recombinant)

Reference Number: PA.CP.PHAR.218

Effective Date: 01/18

Last Review Date: 07/18

[Coding Implications](#)

[Revision Log](#)

Description

The intent of the criteria is to ensure that patients follow selection elements established by Pennsylvania Health and Wellness[®] clinical policy for factor IX* (AlphaNine SD[®], Alprolix[®], BeneFIX[®], Ixinity[®], Mononine[®], Rixubis[®]).

**Factor IX products should not be confused with factor IX complex products (containing factors IX, II, X and VII).*

FDA Approved Indication(s)

AlphaNine SD and Mononine are indicated for the prevention and control of bleeding in patients with Factor IX deficiency due to hemophilia B, also known as Christmas disease.

Limitation(s) of use:

- AlphaNine SD and Mononine contain low, non-therapeutic levels of Factors II, VII, and X, and, therefore, are not indicated for the treatment of Factor II, VII or X deficiencies.
- These products are also not indicated for the reversal of coumarin anticoagulant-induced hemorrhage, nor in the treatment of hemophilia A patients with inhibitors to Factor VIII.
- Mononine is also not indicated in a hemorrhagic state caused by hepatitis-induced lack of production of liver dependent coagulation factors.

Alprolix, Idelvion, and Rixubis are indicated in adults and children with hemophilia B (congenital Factor IX deficiency) for:

- On-demand treatment and control of bleeding episodes;
- Perioperative management of bleeding;
- Routine prophylaxis to reduce the frequency of bleeding episodes.

Limitation(s) of use:

- Alprolix, Idelvion, and Rixubis are not indicated for induction of immune tolerance in patients with hemophilia B.

BeneFIX is indicated in:

- Adult and pediatric patients with hemophilia B (congenital factor IX deficiency or Christmas disease) for
 - Control and prevention of bleeding episodes;
 - Perioperative management.

Limitation(s) of use: BeneFIX is NOT indicated for:

- Treatment of other factor deficiencies (e.g., factors II, VII, VIII, and X);
- Treatment of hemophilia A patients with inhibitors to factor VIII;
- Reversal of coumarin-induced anticoagulation;
- Treatment of bleeding due to low levels of liver-dependent coagulation factors.

Ixinity is indicated in:

- Adults and children ≥ 12 years of age with hemophilia B for:

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- Control and prevention of bleeding episodes;
- Perioperative management.

Limitation(s) of use:

- Ixinity is not indicated for induction of immune tolerance in patients with hemophilia B.

Policy/Criteria

It is the policy of health plans affiliated with Centene Corporation® that AlphaNine SD, Alprolix, BeneFIX, Idelvion, Ixinity, Mononine, Rixubis are **medically necessary** when the following criteria are met:

I. Initial Approval Criteria

A. Congenital Hemophilia B (must meet all):

1. Diagnosis of congenital hemophilia B (factor IX deficiency);
2. Prescribed by or in consultation with a hematologist;
3. If Ixinity is prescribed, age \geq 12 years;
4. If AlphaNine is prescribed, age \geq 17 years;
5. Request is for any of the following:
 - a. Control and prevention of bleeding episodes;
 - b. Perioperative management;
 - c. Routine prophylaxis to prevent or reduce the frequency of bleeding episodes (Alprolix, Idelvion, or Rixubis only);
6. Dose does not exceed the FDA approved maximum recommended dose for the relevant indication.

Approval duration:

3 months (bleeding episodes/surgery)

6 months (routine prophylaxis)

B. Other diagnoses/indications: Refer to PA.CP.PMN.53

II. Continued Approval

A. Congenital Hemophilia B (must meet all):

1. Currently receiving medication via Pennsylvania Health and Wellness benefit or member has previously met all initial approval criteria; or the Continuity of Care policy (PA.LTSS.PHAR.01) applies;
2. Member has responded positively to therapy.
3. If request is for a dose increase, new dose does not exceed the FDA approved maximum recommended dose for the relevant indication.

Approval duration:

3 months (bleeding episodes/surgery)

6 months (routine prophylaxis)

B. Other diagnoses/indications (1 or 2):

1. Currently receiving medication via Pennsylvania Health and Wellness benefit and documentation supports positive response to therapy; or the Continuity of Care policy (PA.LTSS.PHAR.01) applies.

Approval duration: Duration of request or 6 months (whichever is less); or

2. Refer to PA.CP.PMN.53

Background

Description/Mechanism of Action:

Policy products replace deficient clotting factor IX. Factor IX is a vitamin K-dependent coagulation factor which is synthesized in the liver. Factor IX is activated by factor XIa in the intrinsic coagulation pathway. Activated factor IX (IXa) in combination with factor VII activates factor X to Xa, resulting in the conversion of prothrombin to thrombin and the formation of a fibrin clot. Hemophilia B is an X-linked inherited disorder of blood coagulation characterized by insufficient or abnormal synthesis of the clotting protein factor IX. The infusion of exogenous factor IX to replace the deficiency present in hemophilia B temporarily restores hemostasis.

Formulations:

Recombinant:

- Factor IX concentrate with Fc fusion
 - Solution Reconstituted, Intravenous [preservative free]:
 - Alprolix: 250 units; 500 units; 1000 units; 2000 units; 3000 units; 4000 units
- Factor IX concentrate
 - Kit, Intravenous [preservative free]:
 - BeneFIX: 250 units, 500 units, 1000 units, 2000 units, 3000 units
 - Solution Reconstituted, Intravenous [preservative free]:
 - Ixinity: 250 units; 500 units; 1000 units; 1500 units; 2000 units; 3000 units
 - Rixubis: 250 units; 500 units; 1000 units; 2000 units; 3000 units

From human plasma:

- Factor IX concentrate
 - Solution Reconstituted, Intravenous [preservative free]:
 - AlphaNine SD: 500 units; 1000 units; 1500 units
 - Mononine: 1000 units

Appendices

Appendix A: Abbreviation Key

DIC: disseminated intravascular coagulation

Coding Implications

Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

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HCPCS Codes	Description
J7194	Factor IX complex, per IU
J7195	Injection, factor IX (antihemophilic factor, recombinant) per IU, not otherwise specified
J7200	Injection, factor IX, (antihemophilic factor, recombinant), Rixubis, per IU
J7201	Injection, factor IX, FC fusion protein (recombinant), per IU
J7202	Injection, factor IX, albumin fusion protein, (recombinant), Idelvion, per IU.

Reviews, Revisions, and Approvals	Date	Approval Date
Added Idelvion to the policy under the same coverage criteria as the other recombinant factor IX agents. Specified routine prophylaxis indication is only for certain agents, per package labeling for those agents. Added age limit for AlphaNine per package labeling. References reviewed and updated.	02/18	

References

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2. Alprolix Prescribing Information. Cambridge, MA: Biogen Idec, Inc.; November 2017. Available at: www.alprolix.com. Accessed November 28, 2017.
3. BeneFix Prescribing Information. Philadelphia, PA: Wyeth Pharmaceuticals, Inc.; June 2017. Available at: www.benefix.com. Accessed November 28, 2017.
4. Idelvion Prescribing Information. Kankakee, IL: CSL Behring LLC; February 2017. Available at: www.idelvion.com. Accessed November 28, 2017.
5. Ixinity Prescribing Information. Berwyn, PA: Aptevo BioTherapeutics LLC; August 2016. Available at: www.ixinity.com. Accessed November 28, 2017.
6. Mononine Prescribing Information. Kankakee, IL: ZLB Behring, LLC; April 2016. Available at: <http://labeling.cslbehring.com/PI/US/Mononine/EN/Mononine-Prescribing-Information.pdf>. Accessed November 28, 2017.
7. Rixubis Prescribing Information. Westlake Village, CA: Baxalta US Inc.; March 2016. Available at: <http://www.rixubis.com>. Accessed November 28, 2017.
8. Srivastava A, Brewer AK, Mauser-Bunschoten EP, et al. Guidelines for the management of hemophilia. *Haemophilia*. Jan 2013; 19(1): e1-47.
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