Clinical Policy: Factor IX (Human - AlphaNine SD, Mononine; Recombinant - Alprolix, BeneFIX, Idelvion, Ixinity, Rixubis)
Reference Number: ERX.SPMN.200
Effective Date: 01/17
Last Review Date:

See Important Reminder at the end of this policy for important regulatory and legal information.

Policy/Criteria
It is the policy of health plans affiliated with Envolve Pharmacy Solutions™ that factor IX (Human - AlphaNine SD®, Mononine®, Recombinant - Alprolix®, BeneFIX®, Idelvion®, Ixinity®, Rixubis®) is medically necessary when the following criteria are met:

I. Initial Approval Criteria
   A. Hemophilia B (must meet all):
      1. Prescribed by or in consultation with a hematologist;
      2. Diagnosis of hemophilia B;
      3. Prescribed agent will be used for any of the following:
         a. Control and prevention of bleeding episodes;
         b. Perioperative management;
         c. Prophylaxis to control bleeding episodes (Alprolix, Idelvion, or Rixubis only).

      Approval duration: 3 months (bleeding episodes/surgery)
                      6 months (prophylaxis)

   B. Other diagnoses/indications:
      Refer to ERX.SPMN.16 - Global Biopharm Policy.

II. Continued Approval
   A. Hemophilia B (must meet all):
      1. Currently receiving medication via health plan benefit or member has previously met all initial approval criteria.

      Approval duration: 3 months (bleeding episodes/surgery)
                      6 months (prophylaxis)

   B. Other diagnoses/indications (must meet 1 or 2):
      1. Currently receiving medication via health plan benefit and documentation supports positive response to therapy; or
      2. Refer to ERX.SPMN.16 - Global Biopharm Policy.

Background
Description/Mechanism of Action:
Policy products replace deficient clotting factor IX. Hemophilia B, or Christmas disease, is an X-linked inherited disorder of blood coagulation characterized by insufficient or abnormal synthesis of the clotting protein factor IX. Factor IX is a vitamin K-dependent coagulation factor which is synthesized in the liver. Factor IX is activated by factor Xla in the intrinsic coagulation
Clinical Policy
Factor IX

pathway. Activated factor IX (IXa) in combination with factor VII:C activates factor X to Xa, resulting ultimately in the conversion of prothrombin to thrombin and the formation of a fibrin clot. The infusion of exogenous factor IX to replace the deficiency present in hemophilia B temporarily restores hemostasis.

Formulations:
Solution Reconstituted, Intravenous:
  Factor IX, recombinant human with Fc fusion:
    Alprolix (FIXFc Fusion/longer lasting)
  Factor IX, recombinant human without Fc fusion:
    BeneFIX
    Ixinity
    Rixubis
  Factor IX, concentrate from human plasma:
    AlphaNine SD
    Mononine
  Factor IX, recombinant, albumin fusion protein:
    Idelvion

FDA Approved Indications:
AlphaNine SD is indicated for:
  • Prevention and control of bleeding in patients with Factor IX deficiency due to hemophilia B.

Limitations of use:
  • AlphaNine SD contains low, non-therapeutic levels of Factors II, VII, and X, and, therefore, is not indicated for the treatment of Factor II, VII or X deficiencies;
  • This product is also not indicated for the reversal of coumarin anticoagulant-induced hemorrhage, nor in the treatment of hemophilia A patients with inhibitors to Factor VIII.

Alprolix is indicated in:
  • Adults and children with hemophilia B (congenital Factor IX deficiency) for:
    o On demand treatment and control of bleeding episodes;
    o Perioperative management of bleeding;
    o Routine prophylaxis to reduce the frequency of bleeding episodes.

Limitations of use:
  • Alprolix is 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
    o Control and prevention of bleeding episodes;
    o Peri-operative management.

Limitations of use: BeneFIX is NOT indicated for:
  • Treatment of other factor deficiencies (e.g., factors II, VII, VIII, and X);
CLINICAL POLICY
Factor IX

- 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.

Idelvion is indicated in:
- On-demand control and prevention of bleeding episodes
- Perioperative management of bleeding
- Routine prophylaxis to prevent or reduce the frequency of bleeding episodes

Limitations of use:
- Idelvion is not indicated for induction of immune tolerance in patients with hemophilia B.

Ixinity is indicated in:
- Adults and children ≥ 12 years of age with hemophilia B for:
  - Control and prevention of bleeding episodes and for perioperative management.

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

Mononine is indicated for:
- Prevention and control of bleeding in Factor IX deficiency, also known as Hemophilia B or Christmas disease.

Limitations of use:
- Mononine is not indicated in the treatment or prophylaxis of Hemophilia A patients with inhibitors to Factor VIII;
- Mononine contains non-detectable levels of Factors II, VII and X (<0.0025 IU per Factor IX unit using standard coagulation assays) and is, therefore, not indicated for replacement therapy of these clotting factors;
- Mononine is also not indicated in the treatment or reversal of coumarin-induced anticoagulation or in a hemorrhagic state caused by hepatitis-induced lack of production of liver dependent coagulation factors.

Rixubis is indicated in adults and children with hemophilia B for:
- Control and prevention of bleeding episodes;
- Perioperative management;
- Routine prophylaxis.

Limitations of use:
- Rixubis is not indicated for induction of immune tolerance in patients with hemophilia B.

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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<thead>
<tr>
<th>HCPCS Codes</th>
<th>Description</th>
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<tr>
<td>J7194</td>
<td>Factor IX complex, per IU</td>
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<tr>
<td>J7195</td>
<td>Injection, factor IX (antihemophilic factor, recombinant) per IU, not otherwise specified</td>
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<tr>
<td>J7200</td>
<td>Injection, factor IX, (antihemophilic factor, recombinant), Rixubis, per IU</td>
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<tr>
<td>J7201</td>
<td>Injection, factor IX, FC fusion protein (recombinant), per IU</td>
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Reviews, Revisions, and Approvals

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<th>Policy created.</th>
<th>Date</th>
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<td>12/16</td>
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References


Important Reminder

This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of
Factor IX

medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical policy; and other available clinical information.

This Clinical Policy is not intended to dictate to providers how to practice medicine, nor does it constitute a contract or guarantee regarding payment or results. Providers are expected to exercise professional medical judgment in providing the most appropriate care, and are solely responsible for the medical advice and treatment of members.

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