

Clinical Policy: Factor IX Complex (Human - Bebulin, Profilnine)

Reference Number: PA.CP.PHAR.219

Effective Date: 01/18

Last Review Date: 07/18

[Coding Implications](#)

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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 complex* (Bebulin[®], Profilnine[®]).

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

FDA Approved Indication(s)

Bebulin is indicated for:

- Prevention and control of bleeding episodes in adult patients with hemophilia B (congenital factor IX deficiency or Christmas disease).

Limitation(s) of use: Bebulin is not indicated for use in the treatment of factor VII deficiency. No clinical studies have been conducted to show benefit from this product for treating deficiencies other than factor IX deficiency.

Profilnine is indicated for:

- Prevention and control of bleeding in patients with factor IX deficiency (hemophilia B).

Limitation(s) of use: Profilnine contains non-therapeutic levels of factor VII and is not indicated for use in the treatment of factor VII deficiency.

Policy/Criteria

It is the policy of Pennsylvania Health and Wellness that Bebulin and Profilnine 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. Age \geq 18 years;
4. Request is for control and prevention of bleeding episodes;
5. Dose does not exceed the FDA approved maximum recommended dose for the relevant indications.

Approval duration: 3 months

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 is responding 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 indications.

Approval duration: 3 months

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:

Bebulin and Profilnine contain coagulation factors IX, II, X and low levels of factor VII. 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 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. Hemophilia B is an X-linked recessively inherited disorder of blood coagulation characterized by insufficient or abnormal synthesis of the clotting protein factor IX.

Formulations (from human plasma):

Solution Reconstituted, Intravenous:

- Bebulin: 200-1200 (units of factor IX activity stated on each vial)
- Profilnine: 500; 1000; 1500 (units of factor IX activity)

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.

HCPCS Codes	Description
J7194	Factor IX complex, per IU

Reviews, Revisions, and Approvals	Date	Approval Date
Changed age limit for Profilnine to 18 years, per PI. References reviewed and updated	02/18	

References

1. Bebulin Prescribing Information. Westlake Village, CA: Baxter Healthcare Corporation; September 2015. Available at http://www.shirecontent.com/PI/PDFs/BEBULIN_USA_ENG.pdf. Accessed November 28, 2017.
2. Profilnine Prescribing Information. Los Angeles, CA: Grifols Biologicals, Inc.; May 2014. Available at <http://www.grifolsusa.com/documents/10192/89476/ft-profilnine-us-en/03a3eed9-2e02-4e7f-ae7b-9bff623d8535>. Accessed November 28, 2017.
3. Srivastava A, Brewer AK, Mauser-Bunschoten EP, et al. Guidelines for the management of hemophilia. *Haemophilia*. Jan 2013; 19(1): e1-47.
4. Medical and Scientific Advisory Council (MASAC) of the National Hemophilia Foundation (NHF): Database of treatment guidelines. Available at <https://www.hemophilia.org/Researchers-Healthcare-Providers/Medical-and-Scientific-Advisory-Council-MASAC/MASAC-Recommendations>. Accessed November 28, 2017.