CVS/caremark™

SPECIALTY GUIDELINE MANAGEMENT

BEBULIN, PROFILNINE
(factor IX complex [human])

POLICY

A. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication

- Hemophilia B

Compendial Uses

- Bleeding due to low levels of liver-dependent coagulation factors
- Factor X deficiency (Bebulin only)
- Factor II deficiency (Profilnine only)

All other indications are considered experimental/investigational and are not a covered benefit.

B. REQUIRED DOCUMENTATION

The following information is necessary to initiate the prior authorization review:

- Laboratory documentation of the most recent Bethesda titer in members with hemophilia B and inhibitors to factor IX

C. CRITERIA FOR APPROVAL

1. **Hemophilia B**
   Authorization for 12 months may be granted to members prescribed Bebulin or Profilnine for hemophilia B when EITHER of the following criteria is met:
   a. Member has no inhibitors to factor IX
   b. Member has inhibitors to factor IX (see Appendix) and the inhibitor titer is <5 Bethesda units per milliliter[5,8]

2. **Bleeding Due to Low Levels of Liver-dependent Coagulation Factors**
   Authorization for 12 months may be granted to members prescribed Bebulin or Profilnine for bleeding due to low levels of liver-dependent coagulation factors.

3. **Factor X Deficiency**
   Authorization for 12 months may be granted to members prescribed Bebulin for factor X deficiency.[3,5-7]

4. **Factor II Deficiency**
   Authorization for 12 months may be granted to members prescribed Profilnine for factor II deficiency.[3,5,6]

D. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet ALL initial authorization criteria.

E. DOSAGE AND ADMINISTRATION

Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines.

F. APPENDIX

Appendix: Inhibitors - Bethesda Units (BU)

The presence of inhibitors is confirmed by a specific blood test called the Bethesda inhibitor assay.

- High-titer inhibitors:
  - ≥ 5 BU/mL
  - Inhibitors act strongly and quickly neutralize factor
- Low-titer inhibitors:
  - < 5 BU/mL
  - Inhibitors act weakly and slowly neutralize factor

REFERENCES

1. Bebulin [package insert]. Westlake Village, CA: Baxter Healthcare Corporation; July 2012.
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3. Micromedex Solutions [database online]. Ann Arbor, MI: Truven Health Analytics Inc. Updated periodically. www.micromedexsolutions.com [available with subscription]. Accessed December 18, 2015.
4. Clinical Consult: CVS Caremark Clinical Programs Review. Focus on Hemophilia Agents; November 2006.
5. National Hemophilia Foundation. MASAC recommendations concerning products licensed for the treatment of hemophilia and other bleeding disorders. Revised August 2015. MASAC Document # 237. Accessed December 22, 2015.
6. Clinical Consult. CVS Caremark. Clinical Programs Review. Focus on Bleeding Disorder Programs; June 2014.
7. Brown DL, Kouides PA. Diagnosis and treatment of inherited factor X deficiency. *Haemophilia*. 2008;14:1176-1182.
8. *Guidelines for the Management of Hemophilia*. Montreal, Canada: World Federation of Hemophilia, 2012. http://www1.wfh.org/publications/files/pdf-1472.pdf. Accessed December 22, 2015.