

Reference number(s)
1947-A

## SPECIALTY GUIDELINE MANAGEMENT

### NOVOSEVEN RT (coagulation factor VIIa [recombinant])

#### POLICY

##### I. 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.

###### A. FDA-Approved Indications

1. Hemophilia A or hemophilia B with inhibitors
2. Congenital factor VII deficiency
3. Glanzmann's thrombasthenia
4. Acquired hemophilia

###### B. Compendial Uses

1. Acquired von Willebrand syndrome
2. Inhibitors to factor XI

All other indications are considered experimental/investigational and not medically necessary.

##### II. PRESCRIBER SPECIALTIES

Must be prescribed by or in consultation with a hematologist.

##### III. CRITERIA FOR INITIAL APPROVAL

###### A. **Congenital Factor VII Deficiency**

Authorization of 12 months may be granted for treatment of congenital factor VII deficiency.

###### B. **Hemophilia A with Inhibitors**

Authorization of 12 months may be granted for treatment of hemophilia A with inhibitors (see Appendix) when the inhibitor titer is  $\geq 5$  Bethesda units per milliliter (BU/mL) or the member has a history of an inhibitor titer  $\geq 5$  BU.

###### C. **Hemophilia B with Inhibitors**

Authorization of 12 months may be granted for treatment of hemophilia B with inhibitors (see Appendix) when the inhibitor titer is  $\geq 5$  Bethesda units per milliliter (BU/mL) or the member has a history of an inhibitor titer  $\geq 5$  BU.

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**D. Glanzmann's Thrombasthenia**

Authorization of 12 months may be granted for treatment of Glanzmann's thrombasthenia.

**E. Acquired Hemophilia**

Authorization of 12 months may be granted for treatment of acquired hemophilia.

**F. Acquired von Willebrand Syndrome**

Authorization of 12 months may be granted for treatment of acquired von Willebrand syndrome when other therapies failed to control the member's condition (e.g., desmopressin or factor VIII/von Willebrand factor).

**G. Inhibitors to Factor XI**

Authorization of 12 months may be granted for treatment of inhibitors to factor XI.

**IV. CONTINUATION OF THERAPY**

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization for an indication listed in Section III when the member is experiencing benefit from therapy (e.g., reduced frequency or severity of bleeds).

**V. 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:
  - $\geq 5$  BU/mL
  - Inhibitors act strongly and quickly neutralize factor
- Low-titer inhibitors:
  - $< 5$  BU/mL
  - Inhibitors act weakly and slowly neutralize factor

**VI. REFERENCES**

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