Intent:

The intent of this policy/guideline is to provide information to the prescribing practitioner outlining the coverage criteria for Novoseven RT under the patient's prescription drug benefit.

Description:

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.

Applicable Drug List:

Novoseven RT

Policy/Guideline:

Prescriber Specialty:

Must be prescribed by or in consultation with a hematologist.

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 ≥ 5 Bethesda units per milliliter (BU/mL) or the member has a history of an inhibitor titer ≥ 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 ≥ 5 Bethesda units per milliliter (BU/mL) or the member has a history of an inhibitor titer ≥ 5 BU.

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.

Continuation of Therapy:

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

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

Approval Duration and Quantity Restrictions:

Approval: 12 months

References:

1. NovoSeven RT [package insert]. Plainsboro, NJ: Novo Nordisk Inc.; July 2020.
2. National Institutes of Health. The diagnosis, evaluation, and management of von Willebrand disease. Bethesda, MD: US Dept of Health and Human Services, National Institutes of Health; 2007. NIH publication No. 08-5832
3. Tiede A, Rand J, Budde U, et al. How I treat the acquired von Willebrand syndrome. Blood. 2011;117(25):6777-85.
4. Federici AB, Budde U, Castaman G, Rand JH, Tiede A. Current diagnostic and therapeutic approaches to patients with acquired von Willebrand syndrome: a 2013 update. Semin Thromb Hemost. 2013;39(2):191-201.
5. O'Connell NM. Factor XI deficiency – from molecular genetics to clinical management. Blood Coagul Fibrinolysis. 2003;14(Suppl 1):S59-S64.
6. Salomon O, Zivelin A, Livnat T, Seligsohn U. Inhibitors to factor XI in patients with severe factor XI deficiency. Semin Hematol. 2006;43(1 Suppl 1):S10-S12.
7. Srivastava A, Santagostino E, Dougall A, et al. WFH Guidelines for the Management of Hemophilia, 3rd edition. Haemophilia. 2020;26 Suppl 6:1-158. doi:10.1111/hae.14046.
8. National Hemophilia Foundation. MASAC Recommendations Concerning Products Licensed for the Treatment of Hemophilia and Selected Disorders of the Coagulation System. Revised October 2024. MASAC Document #290. https://www.hemophilia.org/sites/default/files/document/files/MASAC-Products-Licensed.pdf. Accessed December 3, 2024.
9. World Federation of Hemophilia. What are inherited platelet function disorders? http://www1.wfh.org/publication/files/pdf-1336.pdf. 2010. Accessed December 3, 2024.
10. World Federation of Hemophilia. Platelet function disorders. http://www1.wfh.org/publication/files/pdf-1147.pdf. 2008. Accessed December 3, 2024.
11. Rajpurkar M, Chitlur M, Recht M, Cooper DL. Use of recombinant activated factor VII in patients with Glanzmann's thrombasthenia: a review of the literature. Haemophilia. 2014;20(4):464-471.
12. Duga S, Salomon O. Congenital factor XI deficiency: an update. Semin Thromb Hemost. 2013;39(6):621-631.