Hemophilia Products – Factor VIIa: NovoSeven RT ™; Sevenfact ® (Intravenous)

Effective Date: 01/01/2020

Review date: 10/02/2019, 12/13/2019, 1/22/20, 6/1/2020, 6/24/2021, 6/16/2022, 6/22/2023

Scope: Medicaid*, Commercial*, Medicare-Medicaid Plan (MMP)
***(Medication only available on the Medical Benefit)**

I. Length of Authorization

Unless otherwise specified*, the initial authorization will be provided for 3 months and may be renewed.

Note: The cumulative amount of medication the patient has on-hand will be taken into account for authorizations. Up to 5 'on-hand' doses for the treatment of acute bleeding episodes will be permitted at the time of the authorization request.

** Initial and renewal authorization periods may vary by specific covered indication*

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

- NovoSeven RT 1000 mcg vial = 12 vials per 30 days
- NovoSeven RT 2000 mcg vial = 12 vials per 30 days
- NovoSeven RT 5000 mcg vial = 24 vials per 30 days
- NovoSeven RT 8000 mcg vial = 15 vials per 30 days
- Sevenfact 1 mg vial = 48 vials per 30 days
- Sevenfact 5 mg vial = 24 vials per 30 days

B. Max Units (per dose and over time) [HCPCS Unit]:

- 120,000 billable units per 30-day supply

III. Initial Approval Criteria [1,2,3,4,9]

Coverage is provided in the following conditions:

- MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements.

NovoSeven RT ONLY[1]

Hemophilia A (congenital factor VIII deficiency) † Φ

- Diagnosis of congenital factor VIII deficiency has been confirmed by blood coagulation testing; **AND**
- Confirmation patient has acquired inhibitors to Factor VIII; **AND**
- Used as treatment in at least one of the following:

- Treatment and control of acute bleeding episodes (episodic treatment of acute hemorrhage); **OR**
- Perioperative management *(*Authorizations valid for 1 month)*; **OR**
- Routine prophylaxis to prevent or reduce the frequency of bleeding episodes when the following criteria are also met:
  - Patient has at least two documented episodes of spontaneous bleeding into joints; **OR**
  - Patient has documented trial and failure of Immune Tolerance Induction (ITI); **AND**
    - Patient has documented trial and failure or contraindication to Hemlibra

Acquired Hemophilia †

- Diagnosis of acquired hemophilia has been confirmed by blood coagulation testing; **AND**
- Used as treatment for one of the following:
  - Treatment and control of acute bleeding episodes (episodic treatment of acute hemorrhage); **OR**
  - Perioperative management *(*Authorizations valid for 1 month)*

Hemophilia B (congenital factor IX deficiency aka Christmas disease) † Φ

- Diagnosis of congenital factor IX deficiency has been confirmed by blood coagulation testing; **AND**
- Confirmation patient has acquired inhibitors to Factor IX; **AND**
- Used as treatment for one of the following:
  - Treatment and control of acute bleeding episodes (episodic treatment of acute hemorrhage); **OR**
  - Perioperative management *(*Authorizations valid for 1 month)*; **OR**
  - Routine prophylaxis to prevent or reduce the frequency of bleeding episodes when the following criteria are also met
    - Patient has at least two documented episodes of spontaneous bleeding into joints; **OR**
    - Patient has documented trial and failure of Immune Tolerance Induction (ITI)

Congenital Factor VII Deficiency † Φ

- Diagnosis of congenital factor VII deficiency has been confirmed by blood coagulation testing; **AND**
- Used as treatment for one of the following:
  - Treatment and control of acute bleeding episodes (episodic treatment of acute hemorrhage); **OR**
  - Perioperative management *(*Authorizations valid for 1 month)*

Glanzmann's Thrombasthenia † Φ

- Diagnosis of Glanzmann Thrombasthenia has been confirmed by blood coagulation testing; **AND**
- Used as treatment for one of the following:
  - Treatment and control of acute bleeding episodes (episodic treatment of acute hemorrhage); **OR**
  - Perioperative management *(*Authorizations valid for 1 month)*; **AND**
- The use of platelet transfusions is known or suspected to be ineffective or contraindicated

Sevenfact ONLY [2]

Hemophilia A (Congenital Factor VIII Deficiency)/Hemophilia B (Congenital Factor IX Deficiency) † Φ

- Patient is at least 12 years of age ; **AND**
- Diagnosis of congenital factor VIII or IX deficiency has been confirmed by blood coagulation testing; **AND**
- Confirmation patient has Hemophilia A (Factor VIII) inhibitors or Hemophilia B (Factor IX) inhibitors; **AND**
- Used as treatment and control of acute bleeding episodes (episodic treatment of acute hemorrhage); **AND**
- Will not be used for the treatment of Congenital Factor VII Deficiency

† FDA Approved Indication(s); ‡ Compendia Recommended Indication(s); **Φ** Orphan Drug

IV. Dispensing Requirements for Rendering Providers (Hemophilia Management Program)

- Prescriptions cannot be filled without an expressed need from the patient, caregiver or prescribing practitioner. Auto-filling is not allowed.
- Monthly, rendering provider must submit for authorization of dispensing quantity before delivering factor product. Information submitted must include:
  - Original prescription information, requested amount to be dispensed, vial sizes available to be ordered from the manufacturer, and patient clinical history (including patient product inventory and bleed history)
  - Factor dose should not exceed +1% of the prescribed dose and a maximum of three vials may be dispensed per dose. If unable to provide factor dosing within the required threshold, below the required threshold, the lowest possible dose able to be achieved above +1% should be dispensed. Prescribed dose should not be increased to meet assay management requirements.
- The cumulative amount of medication(s) the patient has on-hand should be taken into account when dispensing factor product. Patients should not have more than 5 extra doses on-hand for the treatment of acute bleeding episodes.
- Dispensing requirements for renderings providers are a part of the hemophilia management program. This information is not meant to replace clinical decision making when initiating or modifying medication therapy and should only be used as a guide.

V. Renewal Criteria [1,2,3,4,9]

Coverage can be renewed based upon the following criteria:

- Patient continues to meet indication-specific relevant criteria identified in section III ; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: hypersensitivity reactions including anaphylaxis(e.g., hives, itching, rash, difficulty breathing, swelling around the

mouth/throat, chest tightness, wheezing, dizziness/fainting, low blood pressure, etc.), ; serious arterial and venous thrombotic events, development of neutralizing antibodies (inhibitors), etc.;.; **AND**

- Any increases in dose must be supported by an acceptable clinical rationale (i.e., weight gain, half-life study results, increase in breakthrough bleeding when patient is fully adherent to therapy, etc.); **AND**
- The cumulative amount of medication(s) the patient has on-hand will be taken into account when authorizing. The authorization will allow up to 5 doses on-hand for the treatment of acute bleeding episodes as needed for the duration of the authorization; **AND**

Treatment and control of acute bleeding episodes

Renewals will be approved for a 6-month authorization period

Perioperative management of bleeding (NovoSeven RT Only)

- Coverage may NOT be renewed

Routine prophylaxis to prevent or reduce the frequency of bleeding episode (NovoSeven RT Only)

- Renewals will be approved for a 12-month authorization period; **AND**
- Patient has demonstrated a beneficial response to therapy (i.e., the frequency of bleeding episodes has decreased from pre-treatment baseline)

VI. Dosage/Administration[1,2,3,4]

Indication	Dose
NovoSeven RT	
Control and prevention of bleeding: Congenital Hemophilia A or B with inhibitors	Hemostatic Administer 90 mcg/kg intravenously every 2 hours, adjustable based on severity of bleeding until hemostasis is achieved, or until the treatment has been judged to be inadequate. Post-Hemostatic Administer 90 mcg/kg intravenously every 3-6 hours after hemostasis is achieved for severe bleeds
Control and prevention of bleeding: Acquired Hemophilia	Administer 70-90 mcg/kg intravenously every 2-3 hours until hemostasis is achieved
Control and prevention of bleeding: Congenital Factor VII deficiency	Administer 15-30 mcg/kg intravenously every 4-6 hours until hemostasis is achieved
Control and prevention of bleeding: Glanzmann's Thrombasthenia	Administer 90 mcg/kg intravenously every 2-6 hours in severe bleeding episodes requiring systemic hemostatic therapy until hemostasis is achieved
Perioperative management Congenital Hemophilia A or B with inhibitors	Minor

Indication	Dose
	Initial: Administer 90 mcg/kg intravenously immediately before surgery, repeat every 2 hours during surgery. Post-Op: Administer 90 mcg/kg intravenously every 2 hours after surgery for 48 hours, then every 2-6 hours until healing has occurred. Major Initial: Administer 90 mcg/kg intravenously immediately before surgery, repeat every 2 hours during surgery. Post-Op: Administer 90 mcg/kg intravenously every 2 hours after surgery for 5 days, then every 4 hours or by continuous infusion, via pump, at 50 mcg/kg/hr until healing occurs.
Perioperative management Acquired Hemophilia	Administer 70-90 mcg/kg intravenously immediately before surgery and every 2-3 hours for the duration of surgery and until hemostasis is achieved
Perioperative management Congenital Factor VII deficiency	Administer 15-30 mcg/kg intravenously immediately before surgery and every 4-6 hours for the duration of surgery and until hemostasis is achieved
Perioperative management Glanzmann's Thrombasthenia	Initial: Administer 90 mcg/kg intravenously immediately before surgery and repeat every 2 hours for the duration of the procedure. Post-Op: Administer 90 mcg/kg intravenously every 2-6 hours to prevent post-operative bleeding
Sevenfact	
Control and treatment of bleeding: Congenital Hemophilia A or B with inhibitors	For Mild or Moderate Bleeds: – Administer 75 mcg/kg intravenously repeated every 3 hours until hemostasis is achieved or – Initial dose of 225 mcg/kg. If hemostasis is not achieved within 9 hours, additional 75 mcg/kg doses may be administered every 3 hours as needed to achieve hemostasis For Severe Bleeds: – Administer 225 mcg/kg intravenously initially, followed if necessary 6 hours later with 75 mcg/kg every 2 hours until hemostasis is achieved.

VII. Billing Code/Availability Information

HCPCS Code & NDC:

Drug	Manufacturer	J-Code	1 Billable Unit Equiv.	Vial Size	NDC
				1 mg	00169-7010

Novoseven RT	Novo Nordisk	J7189	1 mcg	2 mg	00169-7020
				5 mg	00169-7050
				8 mg	00169-7040
Novoseven RT with MixPro package				1 mg	00169-7201
				2 mg	00169-7202
				5 mg	00169-7205
				8 mg	00169-7208
Sevenfact	LFB S.A.	J7212	1 mcg	1 mg	71127-1000
				5 mg	71127-5000

VIII. References

1. NovoSeven RT [package insert]. Bagsvaerd, Denmark; Novo Nordisk; July 2020. Accessed May 2023.
2. Sevenfact [package insert]. Les Ulis, France; LFB S.A., November 2022. Accessed May 2023.
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4. Guidelines for the Management of Hemophilia. 2nd Edition. World Federation of Hemophilia. 2013. Available at: https://www1.wfh.org/publication/files/pdf-1472.pdf. Accessed January 2019.
5. Annual Review of Factor Replacement Products. Oklahoma Health Care Authority Review Board. Updated April 2016. Access January 2019.
6. Graham A1, Jaworski K. Pharmacokinetic analysis of anti-hemophilic factor in the obese patient. Haemophilia. 2014 Mar;20(2):226-9.
7. Croteau SE1, Neufeld EJ. Transition considerations for extended half-life factor products. Haemophilia. 2015 May;21(3):285-8.
8. Mingot-Castellano, et al. Application of Pharmacokinetics Programs in Optimization of Haemostatic Treatment in Severe Hemophilia a Patients: Changes in Consumption, Clinical Outcomes and Quality of Life. Blood. 2014 December; 124 (21).
9. MASAC RECOMMENDATION CONCERNING PROPHYLAXIS. 2016 National Hemophilia Foundation. MASAC Document #241; February 2016. Available at: http://www.hemophilia.org. Accessed January 2019.
10. First Coast Service Options, Inc. Local Coverage Article: Hemophilia Clotting Factors (A56482). Centers for Medicare & Medicaid Services Inc. Updated on 12/06/2019 with effective date 07/01/2019. Accessed January 2020.
11. Palmetto GBA. Local Coverage Article: Billing and Coding: Guidance for Anti-Inhibitor Coagulant Complex (AICC) National Coverage Determination (NCD) 110.3 (A56065). Centers for Medicare & Medicaid Services Inc. Updated on 10/24/2019 with effective date 10/31/2019. Accessed January 2020.
12. Novitas Solutions, Inc. Local Coverage Article: Billing and Coding: Hemophilia Factor Products (A56433). Centers for Medicare & Medicaid Services Inc. Updated on 11/08/2019 with effective date 11/14/2019. Accessed January 2020.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
D66	Hereditary factor VIII deficiency
D67	Hereditary factor IX deficiency
D68.0	Von Willebrand's disease
D68.2	Hereditary deficiency of other clotting factors
D68.311	Acquired hemophilia
D69.1	Qualitative platelet defects

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD), Local Coverage Determinations (LCDs), and Local Coverage Articles (LCAs) may exist and compliance with these policies is required where applicable. They can be found at: https://www.cms.gov/medicare-coverage-database/search.aspx . Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD/LCA):

Jurisdiction(s): N	**NCD/LCD Document (s):** A56482
https://www.cms.gov/medicare-coverage-database/search/article-date-search.aspx?DocID=A56482&bc=gAAAAAAAAAAA	

Jurisdiction(s): J,M	**NCD/LCD Document (s):** A56065
https://www.cms.gov/medicare-coverage-database/search/article-date-search.aspx?DocID=A56065&bc=gAAAAAAAAAAA	

Jurisdiction(s): H,L	**NCD/LCD Document (s):** A56433
https://www.cms.gov/medicare-coverage-database/search/article-date-search.aspx?DocID=A56433&bc=gAAAAAAAAAAA	

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC