

# Berinert® (C1 Esterase Inhibitor, Human) (Intravenous)

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## I. Length of Authorization

Coverage will be provided for 12 weeks and is eligible for renewal.

The cumulative amount of medication(s) the patient has on-hand, indicated for the acute treatment of HAE, will be taken into account when authorizing. The authorization will provide a sufficient quantity in order for the patient to have a cumulative amount of HAE medication(s) on-hand in order to treat up to 4 acute attacks per 4 weeks for the duration of the authorization (unless otherwise specified).

## II. Dosing Limits

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

- Berinert 500 IU single-dose vial: 20 vials every 28 days

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

- 1000 billable units per 28 days

## III. Initial Approval Criteria <sup>1</sup>

Site of care specialty infusion program requirements are met (refer to [Moda Site of Care Policy](#)).

Coverage is provided in the following conditions:

- Patient has had a documented trial with inadequate response, intolerance or contraindication to the use of generic icatibant; **OR**
- Patient is less than 18 years of age

**Universal Criteria** <sup>1,13,20</sup>

- Must be prescribed by, or in consultation with, a specialist in: allergy, immunology, hematology, pulmonology, or medical genetics; **AND**
- Confirmation the patient is avoiding the following possible triggers for HAE attacks:
  - Estrogen-containing oral contraceptive agents **AND** hormone replacement therapy; **AND**
  - Antihypertensive agents containing ACE inhibitors or angiotensin II receptor blockers (ARBs); **AND**
  - Dipeptidyl peptidase IV (DPP-IV) inhibitors (e.g., sitagliptin); **AND**
  - Neprilysin inhibitors (e.g., sacubitril); **AND**

**Treatment of acute abdominal, facial, or laryngeal attacks of Hereditary Angioedema (HAE) † Φ**  
1,13,20,21,22

- Patient has a history of moderate to severe cutaneous attacks (without concomitant urticaria) **OR** abdominal attacks **OR** mild to severe airway swelling attacks of HAE (i.e. debilitating cutaneous/gastrointestinal symptoms **OR** laryngeal/pharyngeal/tongue swelling); **AND**
- Patient has one of the following clinical presentations consistent with a HAE subtype§, which must be confirmed by repeat blood testing (treatment for acute attack should not be delayed for confirmatory testing):

| <b>HAE I (C1-Inhibitor deficiency) §</b> 13,20,21,22  |
|---|
| <ul style="list-style-type: none"> <li>• Low C1 inhibitor (C1-INH) antigenic level (C1-INH antigenic level below the lower limit of normal as defined by the laboratory performing the test); <b>AND</b></li> <li>• Low C4 level (C4 below the lower limit of normal as defined by the laboratory performing the test); <b>AND</b></li> <li>• Low C1-INH functional level (C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test); <b>AND</b> <ul style="list-style-type: none"> <li>○ Patient has a family history of HAE; <b>OR</b></li> <li>○ Acquired angioedema has been ruled out (i.e., patient onset of symptoms occur prior to 30 years of age, normal C1q levels, patient does not have underlying disease such as lymphoma or benign monoclonal gammopathy [MGUS], etc.)</li> </ul> </li> </ul> |
| <b>HAE II (C1-Inhibitor dysfunction) §</b> 20,22  |
| <ul style="list-style-type: none"> <li>• Normal to elevated C1-INH antigenic level; <b>AND</b></li> <li>• Low C4 level (C4 below the lower limit of normal as defined by the laboratory performing the test); <b>AND</b></li> <li>• Low C1-INH functional level (C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test)</li> </ul>   |

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

#### **IV. Renewal Criteria** <sup>1</sup>

Coverage can be renewed based upon the following criteria:

- Patient continues to meet the universal and other indication-specific relevant criteria identified in section III; **AND**

- Significant improvement in severity and duration of attacks have been achieved and sustained; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: severe hypersensitivity reactions, serious thromboembolic events (arterial or venous), etc.; **AND**
- The cumulative amount of medication(s) the patient has on-hand, indicated for the acute treatment of HAE, will be taken into account when authorizing. The authorization will provide a sufficient quantity in order for the patient to have a cumulative amount of HAE medication(s) on-hand in order to treat up to 4 acute attacks per 4 weeks for the duration of the authorization (unless otherwise specified).

## V. Dosage/Administration <sup>1</sup>

| Indication                               | Dose   |
|--|--|
| Acute Hereditary Angioedema (HAE) attack | Administer 20 international units (IU) per kg body weight by intravenous injection.<br><b>**Note:</b> Patients may self-administer Berinert upon recognition of symptoms of an HAE attack after being instructed by their healthcare provider. |

## VI. Billing Code/Availability Information

### HCPCS Code:

- J0597 – Injection, C-1 esterase inhibitor (human), berinert, 10 units; 1 billable unit = 10 units

### NDC:

- Berinert 500 IU single-dose carton kit (containing a single-dose vial of Berinert and a 10 mL vial of Sterile Water for Injection): 63833-0825-xx
- Berinert 500 IU single-dose vial: 63833-0835-xx

## VII. References

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## Appendix 1 – Covered Diagnosis Codes

| ICD-10 | ICD-10 Description               |
|--------|----------------------------------|
| D84.1  | Defects in the complement system |

## 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): N/A

| 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)          |
| H (4 & 7)   | LA, AR, MS, TX, OK, CO, NM             | Novitas Solutions, Inc.                           |
| 8   | MI, IN                                 | Wisconsin Physicians Service Insurance Corp (WPS) |

**Medicare Part B Administrative Contractor (MAC) Jurisdictions**

| <b>Jurisdiction</b> | <b>Applicable State/US Territory</b>  | <b>Contractor</b>                        |
|---------------------|---|--|
| 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 VA) | Novitas Solutions, Inc.                  |
| K (13 & 14)         | NY, CT, MA, RI, VT, ME, NH  | National Government Services, Inc. (NGS) |
| 15                  | KY, OH  | CGS Administrators, LLC                  |