

## **SPECIALTY GUIDELINE MANAGEMENT**

### **BERINERT (C1 esterase inhibitor [human])**

#### **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.

##### FDA-Approved Indication

Treatment of acute abdominal, facial, or laryngeal attacks of hereditary angioedema (HAE) in adult and pediatric patients.

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

##### **II. DOCUMENTATION**

Submission of the following information is necessary to initiate the prior authorization review:

- A. C4 levels and C1 inhibitor functional and antigenic protein levels
- B. F12, angiotensin-1 or plasminogen gene mutation testing, if applicable
- C. Chart notes confirming family history of angioedema, if applicable

##### **III. CRITERIA FOR INITIAL APPROVAL**

Authorization of 12 months may be granted for treatment of hereditary angioedema attacks when the medication will not be used with Firazyr, Kalbitor, or Ruconest and either of the following criteria is met:

- A. Member has C1 inhibitor deficiency or dysfunction as confirmed by laboratory testing:
  1. C1 inhibitor (C1-INH) antigenic level below the lower limit of normal as defined by the laboratory performing the test; or
  2. Normal C1-INH antigenic level and a low C1-INH functional level (functional C1-INH less than 50% or C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test)
- B. Member has normal C1 inhibitor as confirmed by laboratory testing and meets one of the following criteria:
  1. Member has an F12, angiotensin-1, or plasminogen gene mutation as confirmed by genetic testing, or
  2. Member has a documented family history of angioedema and the angioedema was refractory to a trial of high-dose antihistamine (e.g., cetirizine) for at least one month.

##### **IV. CONTINUATION OF THERAPY**

Authorization of 12 months may be granted for continuation of therapy when all of the following criteria are met:

- A. Member meets the criteria for initial approval.
- B. Member has experienced reduction in severity and/or duration of attacks when they use Berinert to treat an acute attack.

## V. REFERENCES

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4. Zuraw BL, Banerji A, Bernstein JA, et al. US Hereditary Angioedema Association Medical Advisory Board 2013 recommendations for the management of hereditary angioedema due to C1 inhibitor deficiency. *J Allergy Clin Immunol: In Practice.* 2013; 1(5): 458-467.
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6. Maurer M, Magerl M, Ansotegui I, et al. The international WAO/EAACI guideline for the management of hereditary angioedema – the 2017 revision and update. *Allergy.* 2018;00:1-22.
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9. Bowen T. Hereditary angioedema: beyond international consensus – circa December 2010 – The Canadian Society of Allergy and Clinical Immunology Dr. David McCourtie Lecture. *Allergy Asthma Clin Immunol.* 2011;7(1):1.
10. Bernstein J. Update on angioedema: Evaluation, diagnosis, and treatment. *Allergy and Asthma Proceedings.* 2011;32(6):408-412.
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12. Farkas H, Martinez-Saguer I, Bork K, et al. International consensus on the diagnosis and management of pediatric patients with hereditary angioedema with C1 inhibitor deficiency. *Allergy.* 2017;72(2):300-313.
13. Henao MP, Kraschnewski J, Kelbel T, Craig T. Diagnosis and screening of patients with hereditary angioedema in primary care. *Therapeutics and Clin Risk Management.* 2016; 12: 701-711.