

## FORMULARY EXCEPTION POLICY

**POLICY:** Berinert® (C1 esterase inhibitor [human] for intravenous [IV] use – CSL Behring)

**DATE REVISED:** 1/14/2019

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**Documentation:** Documentation will be required for patients requesting Berinert where noted in the criteria as **[documentation required]**. Documentation may include, but is not limited to, chart notes, prescription claims records, prescription receipts and/or laboratory data.

### CRITERIA

- 1. Hereditary Angioedema (HAE) Due to C1 Inhibitor (C1-INH) Deficiency [Type I or Type II] – Treatment of Acute Attacks, Initial Therapy.** Approve Berinert for 1 year if the patient meets the following criteria (A, B and C):
  - A. The patient has HAE type I or type II as confirmed by the following criteria (i and ii):
    - i. Patient has low levels of functional C1-INH protein (< 50% of normal) at baseline, as defined by the laboratory reference values **[documentation required]**; AND
    - ii. Patient has lower than normal serum C4 levels at baseline, as defined by the laboratory reference values **[documentation required]**; AND
  - B. The medication is prescribed by or in consultation with an allergist/immunologist or a physician that specializes in the treatment of HAE or related disorders; AND
  - C. The patient meets ONE of the following criteria (i, ii or iii):
    - i. The patient has tried Ruconest **[documentation required]**; OR
    - ii. The patient has a history of at least one laryngeal attack that had been successfully treated with Berinert, as per the prescribing physician; OR
    - iii. The patient is less than 13 years of age: approve if the patient has tried Cinryze, if formulary. If Cinryze is non-formulary, approve if the patient meets A and B above.
- 2. Hereditary Angioedema (HAE) Due to C1 Inhibitor (C1-INH) Deficiency [Type I or Type II] – Prophylaxis, Initial Therapy.** Approve Berinert for 1 year if the patient meets the following criteria (A, B, and C):
  - A. The patient has HAE type I or type II as confirmed by the following diagnostic criteria (i and ii):
    - i. Patient has low levels of functional C1-INH protein (<50 % of normal) at baseline, as defined by the laboratory reference values **[documentation required]**; AND
    - ii. Patient has lower than normal serum C4 levels at baseline, as defined by the laboratory reference values **[documentation required]**; AND
  - B. The medication is prescribed by or in consultation with an allergist/immunologist or a physician that specializes in the treatment of HAE or related disorders; AND
  - C. The patient has tried ONE of Cinryze, Haegarda, or Takhzyro, if one is formulary. If none are formulary, approve if the patient meets A and B above.
- 3. Hereditary Angioedema (HAE) Due to C1 Inhibitor (C1-INH) Deficiency [Type I or Type II] – Patients Currently Receiving Berinert for Prophylaxis or Acute Therapy.** Approve Berinert for 1 year if the patient meets the following criteria (A, B, and C):
  - A. The patient meets ONE of the following criteria (i or ii):
    - i. The patient meets BOTH of the following criteria (a and b):

- a. Patient is currently receiving Berinert for HAE type I or type II prophylaxis [documentation required to confirm HAE type I or type II diagnosis]; AND
- b. According to the prescribing physician, the patient has had a favorable clinical response (e.g., decrease in number of HAE acute attack frequency, decrease in HAE attack severity, decrease in duration of HAE attacks) since initiating Berinert prophylactic therapy compared with baseline (i.e., prior to initiating prophylactic therapy).
- ii. The patient meets BOTH of the following criteria (a and b):
  - a. The patient is currently treating or has treated previous acute HAE type I or type II attacks with Berinert [documentation required to confirm HAE type I or type II diagnosis]; AND
  - b. According to the prescribing physician, the patient has had a favorable clinical response (e.g., decrease in the duration of HAE attacks, quick onset of symptom relief, complete resolutions of symptoms, decrease in HAE acute attack frequency or severity) with Berinert treatment.
- B.** The medication is prescribed by or in consultation with an allergist/immunologist or a physician that specializes in the treatment of HAE or related disorders; AND
- C.** The patient has tried Cinryze, if formulary. If Cinryze is non-formulary, approve if the patient meets A and B above.

**HISTORY**

Type of Revision	Summary of Changes*
New Policy	--