

Berinert® (C1 esterase inhibitor [human])

When requesting Berinert (C1 esterase inhibitor [human]), the individual requiring treatment must be diagnosed with the following FDA-approved indication or approved compendial use and meet the specific coverage guidelines for the covered indications.

FDA-approved Indication

- Berinert is indicated for the treatment of acute attacks of Hereditary Angioedema (HAE).

Approved Off-label Compendial Use

- Prophylaxis against Hereditary Angioedema (HAE) attacks.

Coverage Guidelines

Hereditary Angioedema (HAE) Due to C1 Inhibitor (C1-INH) Deficiency [Type I or Typell] – Prophylaxis and Treatment

The patient must meet **all** of the following criteria for initial authorization:

- The patient has HAE type I or type II as confirmed by the following diagnostic criteria:
 - Patient has low levels of functional C1-INH protein (less than 50% of normal) at baseline, as defined by the laboratory reference values; AND
 - Patient has lower than normal serum C4 levels at baseline, as defined by the laboratory reference values; AND
- 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.

Approval duration: 12 months

The patient must meet **all** of the following criteria for reauthorization:

- The patient is currently receiving Berinert for HAE type I or type II prophylaxis or has previously received Berinert for acute attacks; AND
- According to the prescribing physician, the patient has had a favorable clinical response defined as one of the following:
 - (e.g., decrease in the duration of HAE attacks, quick onset of symptom relief, complete resolution of symptoms, decrease in HAE acute attack frequency or severity) with Berinert treatment; OR
 - (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 initiation prophylactic therapy); AND
- 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.

Approval duration: 12 months

Dosing Recommendation

Approve up to 20 IU/kg intravenously, no more frequently than once daily.

References

1. Berinert® [prescribing information]. Kankakee, IL: CSL Behring; March 2021.
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3. Craig T, Shapiro R, Vegh A, et al. Efficacy and safety of an intravenous C1-inhibitor concentrate for long-term prophylaxis in hereditary angioedema. *Allergy Rhinol (Providence)*. 2017 Mar 1;8(1):13-19.
4. Busse PJ, Christiansen SC, Riedl MA, et al. US HAEA Medical Advisory Board 2020 guidelines for the management of hereditary angioedema. *J Allergy Clin Immunol Pract*. 2021 Jan;9(1):132-150.e3.
5. Betschel S, Badiou J, Binkley K, et al. The International/Canadian Hereditary Angioedema Guideline [published correction appears in *Allergy Asthma Clin Immunol*. 2020 May 6;16:33]. *Allergy Asthma Clin Immunol*. 2019;15:72.
6. Mauer 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;73(8):1575-1596.
7. Riedl MA, Grivcheva-Panovska V, Moldovan D, et al. Recombinant human C1 esterase inhibitor for prophylaxis of hereditary angio-oedema: a phase 2, multicentre, randomised, double-blind, placebo-controlled crossover trial. *Lancet*. 2017;390:1595-1602.

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