

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 and applicable safety criteria 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 HAE attacks

Coverage Guidelines

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

Initial authorization:

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

Reauthorization:

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

- The patient meets BOTH of the following criteria:
 - 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 (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

- 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 initiation prophylactic therapy).
- 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 Guidelines

- The recommended dose is 20 IU/kg IV.

References

1. Berinert® [prescribing information]. Kankakee, IL: CSL Behring LLC; September 2017.
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3. 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. Available at: <https://onlinelibrary.wiley.com/doi/epdf/10.1111/all.13384>. Accessed on September 12, 2018.
4. Zuraw BL, Bork K, Binkley KE, et al. Hereditary angioedema with normal C1 inhibitor function: consensus of an international expert panel. *Allergy Asthma Proc.* 2012;33:S145-S156.
5. Magerl M, Garmeniz AE, Maas C, et al. Hereditary angioedema with normal C1 inhibitor. Update on evaluation and treatment. *Immunol Allergy Clin N Am.* 2017;37:571-584.
6. 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;1:458-467. Available at: <https://haei.org/wp-content/uploads/2015/04/Zuraw-B-L-US-HAEA-MAB-2013-Recommendations.pdf>. Accessed on September 13, 2018.
7. Zuraw BL, Bernstein JA, Lang DM. A focused parameter update: Hereditary angioedema, acquired C1 inhibitor deficiency, and angiotensin-converting enzyme inhibitor-associated angioedema. *J Allergy Clin Immunol.* 2013;131(6):1491-1493.e25.
8. Zuraw BL. Hereditary angioedema. *N Engl J Med.* 2008;359:1027-1036.
9. Pharming receives complete response letter from FDA [press release]. Pharming Group NV; September 19, 2018. Available at <https://www.pharming.com/pharming-receives-complete-response-letter-from-fda/>. Accessed on September 21, 2018.