

PHARMACY POLICY STATEMENT

Marketplace

DRUG NAME	Berinert (C1 esterase inhibitor (human))
BENEFIT TYPE	Medical or Pharmacy
STATUS	Prior Authorization Required

Berinert, approved by the FDA in 2009, is a plasma-derived concentrate of C1 Esterase Inhibitor (Human) indicated for the treatment of acute abdominal, facial, or laryngeal hereditary angioedema (HAE) attacks in adult and pediatric patients.

HAE is a rare autosomal dominant disease characterized by episodic unpredictable swelling, which can occur in a variety of anatomic locations. The swelling results from excess production of the vasodilator bradykinin. Attacks may be painful and cause functional impairment but are not associated with pruritis. The most common types of HAE are caused by deficiency (type 1) or dysfunction (type 2) of C1 inhibitor (C1-INH). Type 1 is the most prevalent.

Berinert (C1 esterase inhibitor (human)) will be considered for coverage when the following criteria are met:

Hereditary Angioedema

For **initial** authorization:

1. Medication must be prescribed by or in consultation with an allergist or immunologist; AND
2. Member has a diagnosis of HAE type I or type II confirmed by **both** of the following:
 - a) Low C4 level;
 - b) Low (<50% of normal) C1 inhibitor antigenic and/or functional level; AND
3. Medication is being prescribed for the treatment of **acute** HAE attacks; AND
4. Member has documented trial and failure of or contraindication to icatibant (if 18 years of age or older)
5. Medication is **not** being used in combination with another acute HAE therapy (e.g., Kalbitor, Firazyr, Ruconest).
6. **Dosage allowed/Quantity limit:** 20 International Units (IU) per kg body weight by IV injection.
QL: 8 vials per fill

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:

1. Chart notes must document improvement such as faster time to symptom relief or resolution of attack.

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Berinert (C1 esterase inhibitor (human)) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
08/25/2017	New policy for Berinert created. Criteria for each type of HAE specified. Criteria of documentation of attacks, discontinuation of meds that can cause HAE, and restriction on combinations with other meds for acute attacks were added.
01/15/2021	Updated references. Removed age limit. Removed hematology as specialist. Simplified the diagnostic criteria. Removed specific body locations from indication, per clinical guidelines. Removed log book requirement. Reworded the renewal criteria. Extended initial approval duration to 6 months and renewal to 12 months. Removed statement about causative meds. Deleted monthly quantity limit.
06/29/2022	Transferred to new template. Updated references. Added pharmacy as benefit option. Added trial of icatibant for adults.
06/18/2025	Annual review; no updates.

References:

1. Berinert [package insert]. Kankakee, IL: CSL Behring LLC; 2021.
2. Busse PJ, Christiansen SC, Riedl MA, et al. US HAEA Medical Advisory Board 2020 Guidelines for the Management of Hereditary Angioedema [published online ahead of print, 2020 Sep 6]. *J Allergy Clin Immunol Pract*. 2020;S2213-2198(20)30878-3. doi:10.1016/j.jaip.2020.08.046
3. Craig TJ, Levy RJ, Wasserman RL, et al. Efficacy of human C1 esterase inhibitor concentrate compared with placebo in acute hereditary angioedema attacks. *J Allergy Clin Immunol*. 2009;124(4):801-808. doi:10.1016/j.jaci.2009.07.017
4. 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. Published 2019 Nov 25. doi:10.1186/s13223-019-0376-8
5. Bork K, Bernstein JA, Machnig T, Craig TJ. Efficacy of Different Medical Therapies for the Treatment of Acute Laryngeal Attacks of Hereditary Angioedema due to C1-esterase Inhibitor Deficiency. *J Emerg Med*. 2016;50(4):567-80.e1. doi:10.1016/j.jemermed.2015.11.008
6. Maurer M, Magerl M, Betschel S, et al. The international WAO/EAACI guideline for the management of hereditary angioedema - The 2021 revision and update. *World Allergy Organ J*. 2022;15(3):100627. Published 2022 Apr 7. doi:10.1016/j.waojou.2022.100627

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