Cinryze® (C1 esterase inhibitor [human])

When requesting Cinryze (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

Cinryze is indicated for prophylaxis against Hereditary Angioedema (HAE) attacks.

Approved Off-Label Compendial Use

Treatment of acute Hereditary Angioedema (HAE) attacks.

Coverage Guidelines

Hereditary Angioedema (HAE) Due to C1 Inhibitor (C1-INH) Deficiency - Prophylaxis and Treatment

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

- Has HAE type I or type II as confirmed by the following diagnostic criteria:
  - Has low levels of functional C1-INH protein (less than 50% of normal) at baseline, as defined by the laboratory reference values; AND
  - Has lower than normal serum C4 levels at baseline, as defined by the laboratory reference values;
- 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 individual must meet **all** of the following criteria for reauthorization:
Note: An individual who is currently on therapy but has not been previously approved via eviCore coverage determination is reviewed under criteria for initial therapy.

- Is currently receiving Cinryze for HAE type I or Type II prophylaxis or has previously received Cinryze for acute attacks;
- Has had a favorable clinical response defined as one of the following:
  - Decrease in number of HAE acute attack frequency, decrease in HAE attack severity, decrease in duration of HAE attacks since initiating Cinryze prophylactic therapy compared with baseline (i.e., prior to initiating prophylactic therapy); OR
  - Decrease in the duration of HAE attacks, quick onset of symptom relief, complete resolution of symptoms, decrease in HAE acute attack frequency or severity;
- 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 Recommendations

- In adults and adolescents (≥ 12 years of age), administer 1,000 IU intravenously every 3 or 4 days. Doses up to 2,000 IU (not exceeding 80 IU/kg) every 3 or 4 days may be given based on individual response.

- In children 6 to 11 years of age, administer 500 IU intravenously every 3 or 4 days. Doses up to 1,000 IU every 3 or 4 days may be given based on individual patient response.

References

1. Cinryze® [prescribing information]. Lexington, MA: Takeda; November 2024.
2. Zuraw BL. Hereditary angioedema. *N Engl J Med*. 2008;359:1027-1036.
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. Maurer M, Magerl M, Betschel S, et al. The international WAO/EAACI guideline for the management of hereditary angioedema: the 2021 revision and update. Allergy. 2022;77(7):1961-1990.
6. 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.