

## RX.PA.007.MPC Prophylactic Hereditary Angioedema Products: Cinryze and Haegarda

The purpose of this policy is to define the prior authorization process for C1 Inhibitor [human] products: Cinryze and Haegarda,

- C1 Inhibitor [human] intravenous (Cinryze) is indicated for routine prophylaxis against angioedema attacks in adolescent and adult patients with HAE.
- C1 Inhibitor [human] subcutaneous (Haegarda) is indicated for routine prophylaxis against angioedema attacks in adolescents and adult patients with HAE.

### DEFINITIONS

**Hereditary Angioedema (HAE)** – a rare disorder characterized by recurrent attacks of swelling that may involve the peripheral extremities, abdomen, genitalia, face, oropharynx, or larynx due to low levels of endogenous or functional C1 inhibitor.

**Hereditary Angioedema Specialist** – an allergist/immunologist who demonstrates clinical expertise in HAE through research, publication, referrals/consults.

### PROCEDURE

#### A. Initial Authorization Criteria:

*Must meet all of the criteria listed below:*

- Must be prescribed by or under the direction of an HAE specialist
- Must meet the following age requirements:
  - Cinryze- 6 years and older
  - Haegarda- 6 years and older
- Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc) listed in the FDA approved labeling
- Must be used as prophylactic therapy for the prevention of HAE attacks
- Must have a diagnosis of HAE confirmed by ALL of the following laboratory values on two separate instances (copy of laboratory reports required, must include reference ranges):
  - Low C4 complement level (mg/dL) **AND**
  - Normal C1q complement component level (mg/dL) **AND**

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- C1q complement component level is not required for patients under the age of 18 OR patients whose symptoms began before age 18
  - Low C1 esterase inhibitor antigenic level (mg/dL) **OR** Low C1 esterase inhibitor functional level (percent)
- Must be a candidate for HAE prophylaxis therapy, demonstrating at least one of the following (chart documentation of each attack is required):
  - History of frequent HAE attacks defined as two or more HAE attacks per month
  - History of severe HAE attacks defined as one or more abdominal attacks in the past 12 months
  - History of any attack of the respiratory tract which compromised the airway
- Member must not be concomitantly using medications that may exacerbate hereditary angioedema including:
  - Angiotensin-converting enzyme (ACE) inhibitors
  - Estrogen containing medications
- Must have had a trial and failure, intolerance, or contraindication to an attenuated androgen (e.g., danazol, stanozolol, oxandrolone)
- Provider attests that the medication is not being used in combination with other medications indicated for HAE prophylaxis

**B. Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc) listed in the FDA approved labeling.**

**C. Cinryze and Haegarda will be considered investigational or experimental for any other use and will not be covered.**

**D. Reauthorization Criteria:**

All prior authorization renewals are reviewed on an annual basis to determine the Medical Necessity for continuation of therapy. Authorization may be extended at one-year intervals based upon chart documentation from the prescriber that the member's condition has improved based upon the prescriber's assessment while on therapy.

- MPC Renewal
  - Chart documentation confirming positive response to therapy as evidenced by documented decrease of HAE attacks from baseline
  - Must be prescribed by or under the direction of an HAE specialist
- Renewal from Previous Insurer
  - Members who have received prior approval (from insurer other than MPC), or have been receiving medication samples, should be considered under criterion A (Initial Authorization Criteria)

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- Provider has documentation confirming continued positive response to therapy as evidenced by documented decrease of HAE attacks from baseline

### Limitations:

Length of Authorization (if above criteria met)	
Initial Authorization	Up to 4 months
Reauthorization	Up to 1 year

### HCPCS Codes:

Code	Description
J0598	Injection, C1 esterase inhibitor (human), Cinryze, 10 units

### REFERENCES

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9. Craig T, Reidl M, Dykewicz MS, et al. When is prophylaxis for hereditary angioedema necessary? *Ann Allergy Asthma Immunol* 2009;102:366-372
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11. Banerji A, Riedl M, Bernstein J, et al. Lanadelumab for prevention of attacks in hereditary angioedema: results from the phase 3 help study. *Ann Allergy Asthma Immunol* 2017 Nov; 119(5):S5
12. Riedl MA, Bernstein JA, Craig T, et al. An open-label study to evaluate the long-term safety and efficacy of lanadelumab for prevention of attacks in hereditary angioedema: design of the HELP study extension. *Clin Transl Allergy* 2017 Oct 6; 7:36.
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**REVIEW HISTORY**

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
<i>Annual Review</i>	<i>02/2026</i>
<i>Annual Review</i>	<i>02/2025</i>
<i>Annual Review</i> <i>Change in Non-MPC renewal to renewal from previous insurer</i>	<i>02/2024</i>
<i>Annual review</i>	<i>02/2023</i>
<i>Removal of Takhzyro from the policy. Update to approved age range for Haegarda and Cinryze. Update to reauthorization criteria for MPC vs Non-MPC requests</i>	<i>10/2022</i>
<i>Annual review</i>	<i>02/2022</i>
<i>Addition of dosing requirements and off-label restrictions</i>	<i>12/2021</i>
<i>P&amp;T Review</i>	<i>11/2020</i>