# 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**

        - 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
- Non-MPC Renewal
    - Members who have previously been taking Cinrzye or Haegarda and are requesting a non-MPC renewal should be considered under criterion A

(Initial Authorization Criteria)

- Member has not been receiving medication samples for Cinryze or Haegarda; AND
- 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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13. Craig T, Pursun EA, Bork K, et al. WAO guideline for the management of hereditary angioedema. World Allergy Organ J 2012 Dec; 5(12):182-199.

## REVIEW HISTORY

| DESCRIPTION OF REVIEW / REVISION | DATE APPROVED |
|---|---|
| *Annual review* | *02/2023* |
| *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* | *10/2022* |
| *Annual review* | *02/2022* |
| *Addition of dosing requirements and off-label restrictions* | *12/2021* |
| *P&T Review* | *11/2020* |