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
- 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)

- 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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8. Agostoni A, Aygoren-Pursun E, Binkley KE et al. Hereditary and acquired angioedema: problems and progress – proceedings of the third C1 esterase inhibitor deficiency workshop and beyond. *J Allergy Clin Immunol* 2004;114(3 suppl):S51-S131
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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REVIEW HISTORY

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
Annual Review *Change in Non-MPC renewal to renewal from previous insurer*	*02/2024*
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*