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

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

- 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.
- Lanadelumab-flyo (Takhzyro) is indicated for prophylaxis to prevent attacks of hereditary angioedema (HAE) in patients 12 years of age and older

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 a HAE specialist
- Must meet the following age requirements:
  - Cinryze- 9 years and older
  - Haegarda- 12 years and older
  - Takhzyro- 12 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
- Must have had a trial and failure, intolerance, or contraindication to an attenuated androgen (e.g., danazol, stanozolol, oxandrolone)

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, Haegarda, and Takhzyro 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.

Limitations:

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

If the established criteria are not met, the request is referred to a Medical Director for review, if required for the plan and level of request.

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
10. "A Study to Evaluate the Clinical Efficacy and Safety of Subcutaneously Administered C1-esterase Inhibitor in the Prevention of Hereditary Angioedema"; https://clinicaltrials.gov NCT01912456, June 2017
11. Takhzyro [prescribing information]. Lexington, MA: Shire Pharmaceuticals Corp; 2018.
12. 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
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14. 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.

RECORD RETENTION

Records Retention for Evolent Health documents, regardless of medium, are provided within the Evolent Health records retention policy and as indicated in CORP.028.E Records Retention Policy and Procedure.

REVIEW HISTORY

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