RX.PA.007.CCH 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 hereditary angioedema (HAE).
- C1 Inhibitor [human] subcutaneous (Haegarda) is indicated for routine prophylaxis againstangioedema attacks in adolescents and adult patients with HAE.
- Lanadelumab-flyo (Takhzyro) is indicated for prophylaxis to prevent attacks of HAE inpatients 12 years of age and older.

Acute & Preventative Therapies for HAE			
ACUTE		**PREVENTATIVE**	
Berinert	Kalbitor	Cinryze	Takhzyro
Firazyr	Ruconest	Haegarda	Ruconest (off-label use)
		Orladeyo	

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.

POLICY

It is the policy of the Health Plan to maintain a prior authorization process that promotes appropriate utilization of specific drugs with potential for misuse or limited indications. This process involves a review using Food and Drug Administration (FDA) criteria to make a determination of Medical Necessity, and approval by the Medical Policy Committee.

The drugs, Cinrzye, Haegarda, and Takhzyro are subject to the prior authorization process.

PROCEDURE

Initial Authorization Criteria:

- *Must meet all the criteria listed below:* Must be prescribed by or under the direction of a HAE specialist
- Must meet the following age requirements:
  - Cinryze- 6 years and older
  - Haegarda- 6 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 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
- For Cinryze requests:
  - Must have a documented intolerance, contraindication, or clinical reason not to use Haegarda

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 1 year
Reauthorization	Same as initial

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.

Codes:

CPT Codes / HCPCS Codes / ICD-10 Codes		
Code	Brand	Description
J0593	TAKHZYRO	INJECTION LANADELUMAB-FLYO, 1 MG
J0598	CINRYZE	INJECTION C1 ESTERASE INHIBITOR 10 UNITS
J0599	HAEGARDA	INJECTION C1 ESTERASE INHIBITOR 10 UNITS

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REVIEW HISTORY

DESCRIPTION OF REVIEW / REVISION	DATE APPROVED
Initial review	03/22
Updated approval durations to 1 year	02/23

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.

Disclaimer

CountyCare medical payment and prior authorization policies do not constitute medical advice and are not intended to govern or otherwise influence the practice of medicine. The policies constitute only the reimbursement and coverage guidelines of CountyCare and its affiliated managed care entities. Coverage for services varies for individual members in accordance with the terms and conditions of applicable Certificates of Coverage, Summary Plan Descriptions, or contracts with governing regulatory agencies.

CountyCare reserves the right to review and update the medical payment and prior authorization guidelines in its sole discretion. Notice of such changes, if necessary, shall be provided in accordance with the terms and conditions of provider agreements and any applicable laws or regulations.

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