



EVH Clinical Guideline 5000.CC for Acute Hereditary Angioedema Products

Guideline Number: EVH_CG_5000.CC	<u>Applicable Codes</u>	
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STATEMENT

General Information

- *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.*
- *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.*

Purpose

The purpose of this guideline is to define the prior authorization process for acute hereditary angioedema products.

Scope

This guideline applies to all practitioners who are involved in providing the requested drug. This guideline is specific to the Health Plan’s medical benefit.

Plan Design Summary

Requests for acute hereditary angioedema products are subject to the preferred medical drug list program. This program applies to the products specified in this guideline and only to indications that are FDA-approved for the preferred product (as applicable). Coverage for non-preferred products is provided based on clinical circumstances that would exclude the use of the preferred product and may be based on previous use of a product. The coverage review process will ascertain situations where a clinical exception can be made.

Preferred Product Table	
Preferred	Berinerit [c-1 esterase inhibitor (human), J0597]
Non-Preferred	Firazyr (icatibant, J1744) Kalbitor (ecallantide, J1290) Ruconest [c-1 esterase inhibitor (recombinant), J0596]

INITIAL REVIEW CRITERIA

The request must meet all of the criteria listed below.

- Must be prescribed for the treatment of acute HAE attacks
 - *NOTE: Ruconest’s efficacy has not been established in HAE patients with laryngeal*

attacks

- Must be prescribed by or under the direction of a HAE specialist. A HAE specialist is defined as an allergist/immunologist who demonstrates clinical expertise in HAE through research, publication, referrals/consults.
- Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc.) listed in the FDA approved labeling
- 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)
 - Normal C1q complement component level (mg/dL)
 - C1q complement component level is not required for patients under the age of 18 OR patients whose symptoms began before age 18
 - Either of the following:
 - Low C1 esterase inhibitor antigenic level (mg/dL)
 - Low C1 esterase inhibitor functional level (percent)
- Must meet the following age requirements:
 - Berinert – 5 years or older
 - Kalbitor – 12 years or older
 - Firazyr – 18 years or older
 - Ruconest – 13 years or older
- **For non-preferred requests:** Must have a trial and failure or intolerance of the preferred product or have a contraindication to the preferred product

REAUTHORIZATION CRITERIA

All prior authorization renewals are reviewed on an annual basis to determine the Medical Necessity for continuation of therapy. The request must meet all the criteria listed below:

- Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc.) listed in the FDA approved labeling
- Must have chart note documentation from the prescriber showing that the member's condition responded to the requested therapy (e.g., improvement in HAE attack severity and/or duration)
- **For non-preferred requests:** Must have a trial and failure or intolerance of the preferred product or have a contraindication to the preferred product

APPROVAL DURATIONS

Initial Authorization	Up to 6 months
Reauthorization	Same as initial

CODING AND STANDARDS

Codes

Code	Brand	Description
J0596	RUCONEST	INJ C1 ESTERASE INHIBITOR (RECOMBINANT), 10 UNITS
J0597	BERINERT	INJECTION, C-1 ESTERASE INHIB (HUMAN), BERINERT, 10 UNITS
J1290	KALBITOR	INJECTION ECALLANTIDE, 1 MG
J1744	FIRAZYR	INJECTION ICATIBANT, 1 MG

Applicable Lines of Business

<input type="checkbox"/>	CHIP (Children's Health Insurance Program)
<input type="checkbox"/>	Commercial
<input type="checkbox"/>	Exchange/Marketplace
<input checked="" type="checkbox"/>	Medicaid
<input type="checkbox"/>	Medicare Advantage

BACKGROUND

- Berinert® (C1 esterase inhibitor [human]) is indicated for the treatment of acute abdominal, facial, or laryngeal attacks of HAE in adult and pediatric patients.
- Kalbitor® (ecallantide) is approved for the treatment of acute attacks of HAE in patients aged 12 years or older.
- Firazyr® (icatibant) is indicated for the treatment of acute attacks of HAE in adults aged 18 years or older.

- Ruconest® (C1 esterase inhibitor, [recombinant]) is indicated for the treatment of acute attacks in adult and adolescent 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.

POLICY HISTORY

Date	Summary
January 2026	<ul style="list-style-type: none"> ● Updated initial and reauthorization criteria: added dosing criterion, added Ruconest limitation with laryngeal attacks, and updated Berinert age limit ● Added preferred product table/requirement – Berinert added as preferred product
March 2022	<ul style="list-style-type: none"> ● New Guideline

LEGAL AND COMPLIANCE

Guideline Approval

Committee

Reviewed / Approved by Evolent Administrative Services Medical Policy Committee

Disclaimer

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