RX.PA.001.MPC Acute Hereditary Angioedema Products: Berinert® (C1 Esterase Inhibitor, Human) Kalbitor® (Ecallantide), Firazyr® (Icatibant), and Ruconest® (C1 Esterase Inhibitor, Recombinant)

The purpose of this policy is to define the prior authorization process for Acute Hereditary Angioedema (HAE) Products: Berinert® (C1 esterase inhibitor [human]), Kalbitor® (ecallantide), Firazyr® (icatibant), and Ruconest® (C1 esterase inhibitor, [recombinant]).

- 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 age 12 years or older.
- Firazyr® (icatibant) is indicated for the treatment of acute attacks of HAE in adults age 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.

The drugs, Berinert® (C1 esterase inhibitor [human]), Kalbitor® (ecallantide), Firazyr® (icatibant), and Ruconest® (C1 esterase inhibitor, [recombinant]) are subject to the prior authorization process.

PROCEDURE

A. Initial Authorization Criteria:

Must meet all of the criteria listed below:

- Must be prescribed for the treatment of acute HAE 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 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)

  - 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 have received at least one dose of requested product as treatment for acute HAE attack in the past. Chart documentation indicating patient response and ability to tolerate medication is required.
- Must meet the following age requirements:
  - Berinert – 2 years or older
  - Kalbitor – 12 years or older
  - Firazyr – 18 years or older
  - Ruconest – 13 years or older

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. Acute hereditary angioedema products will be considered investigational or experimental for any other use and will not be covered.

D. <u>Reauthorization Criteria:</u>
All prior authorization renewals are reviewed on an annual basis to determine the Medical Necessity for continuation of therapy. Authorization may be extended at 1-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.

<u>Limitations:</u>

Length of Authorization (if above criteria met)	
Initial Authorization	1 fill
Reauthorization	Up to 1 year
Quantity Level Limit	
Firazyr	3 syringes per month

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:
J0596	Injection, C1 esterase inhibitor (recombinant), Ruconest, 10 units
J0597	Injection, C1 esterase inhibitor (human), Berinert, 10 units
J1290	Injection, ecallantide, 1 mg

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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*