HIGHMARK HEALTH OPTIONS

Updated: 09/2026
DMMA Approved: 10/2026

Request for Prior Authorization for Hereditary Angioedema (HAE) Agents
Website Form – www.highmarkhealthoptions.com
Submit request via: Fax - 1-855-476-4158

All requests for Hereditary Angioedema (HAE) Agents require a Prior Authorization and will be screened for medical necessity and appropriateness using the criteria listed below.

Hereditary Angioedema (HAE) Agents Prior Authorization Criteria:

For all requests for Hereditary Angioedema (HAE) Agent all of the following criteria must be met:

- The medication requested is used for the management of HAE
- Is age-appropriate according to FDA-approved package labeling, nationally recognized compendia, or peer-reviewed medical literature
- Medication is being prescribed by an expert physician who is knowledgeable about the condition, has experience in managing patients with Hereditary Angioedema (HAE), and is familiar with all HAE management options (e.g., allergy and immunology specialist, hematologist, or dermatologist)
- The diagnosis of HAE is confirmed by laboratory values obtained on two separate instances (laboratory reports must contain reference ranges):
    - HAE I
        - Low C4 level; AND
        - Low C1-INH antigenic level
    - HAE II
        - Low C4 level; AND
        - Normal or elevated C1-INH antigenic level AND low C1-INH functional level
    - HAE III (with normal C1 inhibitor)
        - Normal C4 complement level (mg/dL); AND
        - Normal C1 esterase inhibitor antigenic level (mg/dL); AND
        - Normal C1 esterase inhibitor functional level; AND
        - Has a history of recurrent angioedema without urticaria
        - One of the following:
            - Documentation of a family history of hereditary angioedema; OR
            - Has a hereditary angioedema-causing genetic mutation
- There is a documented history of at least one symptom of a moderate to severe HAE attack (e.g., moderate to severe abdominal pain, facial swelling, airway swelling) in the absence of hives or a medication known to cause angioedema.
- The member is not taking any medications that may exacerbate HAE, including angiotensin-converting enzyme (ACE) inhibitors and estrogen-containing medications.
- The requested dose and frequency is in accordance with FDA-approved labeling, nationally recognized compendia, and/or evidence-based practice guidelines

Coverage may be provided for the acute treatment of HAE and the following criteria is met:

- Request must be for Firazyr (icatibant), Kalbitor (ecallantide), Berinert (C1 esterase inhibitor [recombinant]), or Ruconest (C1 esterase inhibitor [recombinant]) or Ekterly (sebetralstat)
- The member is receiving no more than one medication for the acute treatment of an HAE attack at one time
- If the member is experiencing multiple HAE attacks per month necessitating an amount of medication over the quantity limit, the member is adherent to prophylactic treatment (or clinically relevant contraindication, inadequate response to, or intolerance to prophylactic therapy is documented by the prescriber). If receiving prophylactic therapy, the member is adherent based upon pharmacy claims or physician attestation.
- Must provide documentation showing the member has tried and failed (which will be verified via pharmacy claims if available) or had an intolerance or contraindication to the preferred agents on the Delaware Preferred Drug List if requesting non-preferred agents
- **Initial Duration of Approval:** 3 months
- **Reauthorization criteria**
    - Member continues to meet coverage criteria for HAE management and treatment
    - Must meet one of the following:
        - If taking an attenuated androgen, documentation the physician has completed a patient assessment at least twice within the past year
        - Documentation the physician has completed a patient assessment at least once within the past year
    - Documentation the member has clinically benefitted from medication in an acute HAE attack
- **Reauthorization Duration of approval:** 3 months

Coverage may be provided for short-term prophylaxis of HAE and the following criteria is met:

- Request must be for Cinryze (C1 esterase inhibitor [human]) or Haegarda (C1 esterase inhibitor [human])
- The member is scheduled to undergo a surgical procedure or major dental work
- The type of procedure and date of procedure must be provided
- There is documentation of therapeutic failure, intolerance or contraindication with Haegarda (C1 esterase inhibitor [human])
- **Duration of Approval:** 1 month

Coverage may be provided for long-term prophylaxis of HAE and the following criteria is met:

- Request must be for Cinryze, Takhzyro (lanadelumab-flyo), Haegarda, Danazol, Orladeyo (berotralstat), Andembry (garadacimab-gxii) or Dawnzera (donidalorsen).
- Member has a history of one or more attacks per month resulting in documented E.R. treatment or hospitalization, a history of laryngeal attacks, two or more attacks per month involving the face, throat, or abdomen, or a history of multiple minor attacks sufficient to require quantities of medication exceeding quantity limits
- There is documentation of therapeutic failure, intolerance or contraindication with Haegarda (C1 esterase inhibitor [human])

- **Initial Duration of Approval:** 6 months
- **Reauthorization criteria**
    - Member continues to meet coverage criteria for HAE management and prophylaxis
    - Must meet one of the following:
        - If taking an attenuated androgen, documentation the physician has completed a patient assessment at least twice within the past year
        - Documentation the physician has completed a patient assessment at least once within the past year
    - Documentation demonstrating disease state improvement (e.g., a decrease in the number, severity, and/or duration of acute HAE attacks) is provided
- **Reauthorization Duration of approval:** 12 months

All requests deemed medically necessary and approved will be referred to care management for assistance with disease state management, avoidance of HAE attack triggers and help with other psychosocial issues.

Coverage may be provided for any non-FDA labeled indication if it is determined that the use is a medically accepted indication supported by nationally recognized pharmacy compendia or peer-reviewed medical literature for treatment of the diagnosis(es) for which it is prescribed. These requests will be reviewed on a case by case basis to determine medical necessity.

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HEREDITARY ANGIOEDEMA ACUTE TREATMENT
PRIOR AUTHORIZATION FORM- Page 1 of 2

Please complete and fax all requested information below including any progress notes, laboratory test results, or chart documentation as applicable to Highmark Health Options Pharmacy Services. **FAX: (855) 476-4158**
If needed, you may call to speak to a Pharmacy Services Representative. **PHONE**: (844) 325-6251 Mon – Fri 8 am to 7 pm

PROVIDER INFORMATION

Requesting Physician:	NPI:
Physician Specialty:	Office Contact:
Office Address:	Office Phone:
	Office Fax:
Pharmacy Name:	Pharmacy Phone:

MEMBER INFORMATION

Member Name:	DOB:	
Member ID:	Member weight:	Height:

DRUG INFORMATION

Medication:	Strength	
Directions:	Quantity:	Refills:
Is the member currently receiving requested medication? ☐ Yes ☐ No	Date Medication Initiated:	

Is this medication being used for a chronic or long-term condition for which the medication may be necessary for the life of the patient? ☐ Yes ☐ No

MEDICAL HISTORY (Please attach applicable lab reports and progress notes)

Diagnosis:

1.) Hereditary angioedema ☐ Type 1 ☐ Type 2 ☐ Type 3_____
2.) Has the member's diagnosis been confirmed by lab values obtained on 2 separate instances? ☐ Yes ☐ No
3.) Does the member have a history of at least 1 symptom of a moderate to severe HAE attack? ☐ Yes ☐ No

Is the member receiving more than one medication for the acute treatment of HAE? ☐ Yes ☐ No

Is the member taking any drugs that may exacerbate HAE (ACE inhibitors, estrogen-containing drugs)? ☐ Yes ☐ No

HAE attack history:

How many HAE attacks per month is the member experiencing? _____
What is the location and severity of the HAE attacks? _____

Is the member currently on prophylactic therapy for HAE? ☐ Yes ☐ No
If **yes**, what medication? _____
Has the member been adherent to the prescribed prophylactic regimen? ☐ Yes ☐ No

REAUTHORIZATION

How many doses of acute treatment does the member **currently** have on-hand? _____
How many HAE attacks per month has the member experienced since the last authorization? _____
How many doses of acute treatment have been required per HAE attack? _____
What is the planned frequency of phone assessments and/or office visits? _____
Is the member currently taking an attenuated androgen (e.g. danazol)? ☐ Yes ☐ No
Has the most recent chart documentation or notes been provided? ☐ Yes ☐ No
Has treatment resulted in clinical benefit during an acute HAE attack? ☐ Yes ☐ No
Please describe:_____

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HEREDITARTY ANGIOEDEMA ACUTE TREATMENT PRIOR AUTHORIZATION FORM (CONTINUED) – PAGE 2 OF 2		
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MEMBER INFORMATION		
Member Name:	DOB:	
Member ID:	Member weight:	Height:
SUPPORTING INFORMATION or CLINICAL RATIONALE		
Prescribing Provider Signature	Date	