AvMed

PHARMACY PRIOR AUTHORIZATION/STEP-EDIT REQUEST*

Directions: **The prescribing physician must sign and clearly print name (preprinted stamps not valid) on this request**. All other information may be filled in by office staff; **fax to 1-305-671-0200**. No additional phone calls will be necessary if all information (including phone and fax #s) on this form is correct. **If the information provided is not complete, correct, or legible, the authorization process can be delayed.**

Prophylaxis Hereditary Angioedema (HAE) (Pharmacy)

Drug Requested: (select one drug below)

PREFERRED	
❑ **Cinryze®** (C1 Esterase Inhibitor Human)	❑ **Haegarda®** (C1 Esterase Inhibitor Human)
❑ **Takhzyro®** (lanadelumab)	
NON-PREFERRED	
❑ **Andembry®** (garadacimab-gxii)	❑ **Dawnzera™** (donidalorsen)
❑ **Orladeyo®** (berotralstat)	

MEMBER & PRESCRIBER INFORMATION: Authorization may be delayed if incomplete.

Member Name: _____

Member AvMed #: _____ **Date of Birth:** _____

Prescriber Name: _____

Prescriber Signature: _____ **Date:** _____

Office Contact Name: _____

Phone Number: _____ **Fax Number:** _____

NPI #: _____

DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Form/Strength: _____

Dosing Schedule: _____ **Length of Therapy:** _____

Diagnosis: _____ **ICD Code:** _____

Weight (if applicable): _____ **Date weight obtained:** _____

Dosing Limits: (see below)

- ❑ **Andembry:** Administer initial loading dose of 400 mg (two injections of 200 mg) subcutaneously on the first day of treatment followed by a maintenance dosage of 200 mg every month
  - Quantity Limit: 200 mg auto-injector or syringe: 4 injections per 30 days

(Continued on next page)

- ❑ **Cinryze:** Administer 1,000 units intravenous every 3 or 4 days
  - Quantity Limit: Cinryze 500-unit vial: 16 vials per 30 days
- ❑ **Dawnzera:** 80 mg administered subcutaneously every 4 weeks. A dosage of 80 mg every 8 weeks may also be considered.
  - Quantity Limit: 80 mg auto-injector: 1 injection per 28 days
- ❑ **Haegarda:** Administer 60 International Units (IU) per kg body weight twice weekly (every 3 or 4 days)
  - Quantity Limit:
    - Haegarda 2000 IU SDV kit: 16 kits per 28 days
    - Haegarda 3000 IU SDV kit: 8 kits per 28 days
- ❑ **Orladeyo:** Adults and children ≥ 2 years of age - One capsule/packet taken orally once daily with food
  - Quantity Limit:
    - 150 mg capsules: 1 capsule per day (For Adults and children 12 years of age or older)
    - 110 mg capsules: 1 capsule per day (For members with moderate to severe impairment (Child-Pugh class B and C))
    - 132 mg pellets: 1 packet per day (For members weighing ≥ 40 kg)
    - 108 mg pellets: 1 packet per day (For members weighing 32 to < 40 kg)
    - 96 mg pellets: 1 packet per day (For members weighing 24 to < 32 kg)
    - 72 mg pellets: 1 packet per day (For members weighing 12 to < 24 kg)
- ❑ **Takhzyro:** For children ≥ 12 years of age, adolescents, and adults – Administer 300 mg every 2 weeks. Dosing every 4 weeks for well controlled members (e.g., attack free) for > 6 months. The recommended dosage of Takhzyro in pediatric patients 2 to less than 6 years of age is 150 mg administered SC once every 4 weeks
  - Quantity Limit:
    - 300 mg vial/syringe per 14 days
    - 150 mg syringe per 28 days

CLINICAL CRITERIA: Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

Initial Authorization: 12 months

- ❑ Prescribed by or in consultation with a specialist in allergy, immunology, hematology, pulmonology or medical genetics

AND

- ❑ Member must meet medication specific age requirement:
  - ❑ For Andembry & Dawnzera requests: Member must be at least 12 years of age
  - ❑ For Cinryze & Haegarda requests: Member must be at least 6 years of age
  - ❑ For Orladeyo & Takhzyro requests: Member must be at least 2 years of age

AND

- ❑ Provider attests the member is avoiding **BOTH** of the following possible triggers for HAE attacks:
  - ❑ Estrogen-containing oral contraceptive agents **AND** hormone replacement therapy

(Continued on next page)

- ❑ Antihypertensive agents containing ACE inhibitors

AND

- ❑ Member has a history of **ONE** of the following:
  - ❑ **Three (3) or more** severe HAE attacks per month **(select all that apply)**:
    - ❑ Moderate to severe cutaneous attacks (without concomitant hives)
    - ❑ Abdominal attacks (pain and swelling)
    - ❑ Mild to severe airway swelling attacks of HAE (i.e. laryngeal/pharyngeal/tongue swelling)
  - ❑ Disablement for more than 5 days per month by HAE

AND

- ❑ Provider attests the requested medication will **NOT** be used in combination with other prophylactic therapies targeting C1 inhibitors or kallikrein (e.g., Haegarda or Takhzyro)

AND

- ❑ Treatment of member with "on-demand" therapy (i.e., Berinert, Ekterly, icatibant, Kalbitor or Ruconest) did **NOT** provide satisfactory control or access to "on-demand therapy" is limited **(failure is defined as more than 5 attacks/month for 4 months consecutively within the same year)**

AND

- ❑ Member failed, is intolerant, or has a contraindication to attenuated (17 alpha-alkylated) androgens (e.g., Danazol®) for HAE prophylaxis

AND

- ❑ **For Andembry, Dawnzera & Orladeyo Requests:**
  - ❑ Member has tried and failed **ONE** of the following **(verified by chart notes and/or pharmacy paid claims)**:
    - ❑ Cinryze
    - ❑ Haegarda
    - ❑ Takhzyro

AND

Member has ONE of the following clinical presentations that is consistent with a HAE subtype, confirmed by repeat blood testing (please submit chart notes for symptoms and lab values to confirm the HAE subtype):

II.A. ❑ HAE I: (all bullet points must apply)

- ❑ Low C1 inhibitor (C1-INH) antigenic level (C1-INH antigenic level below the lower limit of normal as defined by the laboratory performing the test)
- ❑ Low C4 level (C4 below the lower limit of normal as defined by the laboratory performing the test)
- ❑ Low C1-INH functional level (C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test) **in addition to ONE** of the following:
  - ❑ Member has a family history of HAE

(Continued on next page)

- ❑ Acquired angioedema has been ruled out (i.e., patient onset of symptoms occur prior to 30 years old, normal C1q levels, patient does not have underlying disease such as lymphoma or benign monoclonal gammopathy [MGUS])

OR

II.B. ❑ HAE II (C1-Inhibitor dysfunction): (all bullet points must apply)

- ❑ Normal to elevated C1-INH antigenic level
- ❑ Low C4 level (C4 below the lower limit of normal as defined by the laboratory performing the test)
- ❑ Low C1-INH functional level (C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test) **in addition to ONE** of the following:
  - ❑ Member has a family history of HAE
  - ❑ Acquired angioedema has been ruled out (i.e., onset of symptoms occurred prior to 30 years old, normal C1q levels, patient does not have underlying disease such as lymphoma or benign monoclonal gammopathy [MGUS])

OR

II.C. ❑ HAE III with normal C1-INH: (all bullet points must apply)

- ❑ Normal C1-INH antigenic level
- ❑ Normal C4 level
- ❑ Normal C1-INH functional level
- ❑ Repeat blood testing during an attack has confirmed the member does **NOT** have abnormal lab values indicative of HAE I or HAE II
- ❑ Member had an inadequate response or intolerance to an adequate trial of prophylactic therapy with **ONE** of following:
  - ❑ antifibrinolytic agent: (❑ tranexamic acid (TXA) **OR** ❑ aminocaproic acid)
  - ❑ 17α- alkylated androgen: danazol
  - ❑ progestins (female members only)

AND

- ❑ **ONE of the following:**
  - ❑ Member has a known HAE-causing mutation (e.g., mutation of coagulation factor XII gene [F12 mutation], mutation in the angiopoietin-1 gene, mutation in the plasminogen gene or kininogen-1)
  - ❑ Member has a family history of HAE and documented evidence of lack of efficacy of chronic high-dose antihistamine therapy (e.g., cetirizine standard dosing at up to four times daily or an alternative equivalent, given for at least one month or an interval long enough to expect three or more angioedema attacks) **AND** corticosteroids

(Continued on next page)

Reauthorization: Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

- ❑ Member must continue to meet initial authorization criteria

AND

- ❑ Significant improvement in severity and duration of attacks has been achieved and sustained

AND

- ❑ Member has experienced an absence of unacceptable toxicity from the drug (e.g., hypersensitivity reactions)

AND

- ❑ For Takhzyro Renewal Only: Has the member been attack free for greater than > 6 months?
  - ❑ Yes- approve dosing every 4 weeks
  - ❑ No- approve dosing every 2 weeks

Medication being provided by a Specialty Pharmacy – Proprium Rx

Use of samples to initiate therapy does not meet step edit/ preauthorization criteria.

Previous therapies will be verified through pharmacy paid claims or submitted chart notes.*