HARVARD PILGRIM HEALTH CARE
RECOMMENDED MEDICATION REQUEST GUIDELINES

HEREDITARY ANGIOEDEMA MEDICATIONS

Generic	Brand	HICL	GCN	Exception/Other
C1 ESTERASE INHIBITOR	CINRYZE HAEGARDA	18568		BRAND ≠ BERINERT
ICATIBANT	FIRAZYR	35962		

GUIDELINES FOR USE

INITIAL CRITERIA (NOTE: FOR RENEWAL CRITERIA SEE BELOW)

1. Is the request for a patient with the diagnosis of Hereditary Angioedema (HAE) and does the request meet **ALL** of the following criteria?
   - Prescribed by, or in consultation with, an allergist, immunologist, or hematologist
   - **ONE** of the following age requirements:
     - 12 years of age or older if the request is for Haegarda
     - 6 years of age or older if the request is for Cinryze
     - 18 years of age or older if the request is for Firazyr
   - Documentation submitted of diagnostic laboratory testing confirming **ONE** of the following conditions:
     - C1 inhibitor deficiency (determined by a C1 inhibitor antigenic protein level or C1 inhibitor functional level below the lower limit of normal) **OR**
     - Normal C1 inhibitor and **each** of the following:
       - Other causes of angioedema have been ruled out **AND**
       - One of the following:
         - Patient tested positive for the F12 gene mutation **OR**
         - Family history of angioedema which was refractory to antihistamine (e.g., cetirizine) therapy

   If yes, continue to #2.
   If no, do not approve. Please use status code #238 and the denial text provided.
   DENIAL TEXT: Per your health plan's **HEREDITARY ANGIOEDEMA MEDICATIONS** guideline, this medication is only covered when prescribed for Hereditary Angioedema (HAE) when you meet ALL of the following conditions:
   - Prescribed by, or in consultation with, an allergist, immunologist, or hematologist
   - An age requirement: **[select based on requested medication]**
     - 12 years of age or older if the request is for Haegarda
     - 6 years of age or older if the request is for Cinryze
     - 18 years of age or older if the request is for Firazyr

   (Denial text continued on next page)

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INITIAL CRITERIA (CONTINUED)

- Documentation submitted of diagnostic laboratory testing confirming **ONE** of the following conditions:
  - C1 inhibitor deficiency (determined by a C1 inhibitor antigenic protein level or C1 inhibitor functional level below the lower limit of normal) **OR**
  - Normal C1 inhibitor and **each** of the following:
    - Other causes of angioedema have been ruled out **AND**
    - One of the following:
      - Patient tested positive for the F12 gene mutation **OR**
      - Family history of angioedema which was refractory to antihistamine (e.g., cetirizine) therapy

Your provider did not submit information that you **[criteria not met]** and therefore your request was not approved.

2. Is the request for Firazyr?

   If yes, **approve for 12 months by HICL for up to a maximum of 6 (3 mL) syringes per Rx.**
   APPROVAL TEXT: Your request for Firazyr has been approved for a quantity up to 6 syringes per prescription for a 12-month period.

   If no, continue to #3.

3. Is there a need for routine prophylaxis as determined by at least **ONE** of the following?
   - One or more HAE attacks per month
   - History of laryngeal attacks
   - Emergency medical care related to HAE three or more times a year

   If yes, continue to #4.
   If no, do not approve. Please use status code #238 and the denial text provided.
   DENIAL TEXT: Per your health plan's **HEREDITARY ANGIOEDEMA MEDICATIONS** guideline, this medication is covered for routine prophylaxis of Hereditary Angioedema (HAE) if you provider submits information that you have a history of one of the following: one or more HAE attacks per month, history of laryngeal attacks, or emergency medical care related to HAE three or more times a year. Your provider did not indicate that you meet the above criteria and therefore your request was not approved.

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HEREDITARY ANGIOEDEMA MEDICATIONS

INITIAL CRITERIA (CONTINUED)

4. Has the patient had an insufficient response, contraindication, or is there clinical rationale for not using androgen therapy (e.g., danazol, oxandrolone, methyltestosterone)?

   If yes, **approve for 12 months by HICL as follows:**
   - For Cinryze, the quantity is hard coded for 20 vials per 30 days. **NOTE**: each single-use vial contains 500 units (USP).
   - For Haegarda, **approve up to the maximum # of vials per 28 days as follows:**

Dosing: 60 IU/kg twice weekly		Max # of 2,000 IU SDVs per 28 days	Max # of 3,000 IU SDVs per 28 days
lbs	kgs		
< 75	< 34	8	
75-100	34-45.5		8
101-150	45.6-68	16	
151-200	68.1-90.9	24	16
201-222	91-100.9	24	
223-295	101-133.9	32	
296-350	134-159	40	

Please use status code #056 and the approval text provided.
APPROVAL TEXT (Cinryze): Your request for Cinryze has been approved for a quantity of 20 vials per 30 days for a 12-month period.

APPROVAL TEXT (Haegarda): Your request for Haegarda has been approved for a quantity up to **[#] [2,000 IU *-or-* 3,000 IU]** vials per 28 days for a 12-month period.

If no, do not approve. Please use status code #238 and the denial text provided.
DENIAL TEXT: Per your health plan's **HEREDITARY ANGIOEDEMA MEDICATIONS** guideline, this medication is only covered for patients with a documented insufficient response, contraindication, or clinical rationale submitted by the provider for not using androgen therapy (such as danazol, oxandrolone, methyltestosterone). Your provider did not indicate that you have failed or you should not use androgen therapy and therefore your request was not approved.

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HARVARD PILGRIM HEALTH CARE
RECOMMENDED MEDICATION REQUEST GUIDELINES

HEREDITARY ANGIOEDEMA MEDICATIONS

RENEWAL CRITERIA

1. Is the medication being prescribed by (or in consultation with) an allergist, immunologist, or hematologist for the diagnosis of Hereditary Angioedema (HAE)?

   If yes, continue to #2.
   If no, do not approve. Please use status code #238 and the denial text provided.
   DENIAL TEXT: Per your health plan's **HEREDITARY ANGIOEDEMA MEDICATIONS** guideline, this medication is only covered for Hereditary Angioedema (HAE) when prescribed by (or in consultation with) an allergist, immunologist, or hematologist. Your provider did not indicate **[criteria not met]** and therefore your request was not approved.

2. Has the patient exhibited a decrease in frequency of HAE attacks **OR** shown improvement in severity and duration of attacks?

   If yes, **approve for 12 months by HICL as follows:**
   - For Cinryze, the quantity is hard coded for 20 vials per 30 days. **NOTE**: each single-use vial contains 500 units.
   - For Haegarda, **approve up to the maximum # of vials per 28 days as follows:**

Dosing: 60 IU/kg twice weekly		Max # of 2,000 IU SDVs per 28 days	Max # of 3,000 IU SDVs per 28 days
lbs	kgs		
< 75	< 34	8	
75-100	34-45.5		8
101-150	45.6-68	16	
151-200	68.1-90.9	24	16
201-222	91-100.9	24	
223-295	101-133.9	32	
296-350	134-159	40	

   - For Firazyr, **approve up to a maximum of 6 (3 mL) syringes per Rx.**

   Please use status code #056 and the approval text provided.
   APPROVAL TEXT (Cinryze): Your request for Cinryze has been approved for a quantity of 20 vials per 30 days for a 12-month period.
   APPROVAL TEXT (Haegarda): Your request for Haegarda has been approved for a quantity up to **[#] [2,000 IU *-or-* 3,000 IU]** vials per 28 days for a 12-month period.
   APPROVAL TEXT (Firazyr): Your request for Firazyr has been approved for a quantity up to 6 syringes per prescription for a 12-month period.

   If no, do not approve. Please use status code #238 and the denial text provided.
   DENIAL TEXT: Per your health plan's **HEREDITARY ANGIOEDEMA MEDICATIONS** guideline, this medication is only covered for patients with a documented decrease in frequency of HAE attacks **OR** documented improvement in severity and duration of attacks. Your provider did not indicate that you meet this condition and therefore your request was not approved.

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HEREDITARY ANGIOEDEMA MEDICATIONS

RATIONALE

To ensure appropriate use of the medications used for the treatment and prophylaxis of Hereditary Angioedema (HAE).

FDA APPROVED INDICATIONS

- Cinryze is indicated for routine prophylaxis against angioedema attacks in adolescent and adult patients with Hereditary Angioedema (HAE).
- Firazyr is indicated for treatment of acute attacks of hereditary angioedema (HAE) in adults 18 years of age and older.
- Haegarda is indicated for routine prophylaxis to prevent Hereditary Angioedema (HAE) attacks in adolescent and adult patients.

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