curative

Hereditary Angioedema (HAE) Prior Authorization

Drug(s) Applied:	**icatibant, Haegarda** (C1 Esterase Inhibitor (Human))

Criteria:
Drug(s) Applied will be approved when the requested medication is being used for an FDA approved indication and all of the following criteria are met:

I. Initial Therapy Criteria
- **A. Hereditary angioedema, acute treatment** as indicated by chart notes within past 120 days
  1. Requested drug is icatibant and will be used to treat acute HAE attacks **and**
  2. ONE of the following:
     a) Diagnosis of C1INH deficiency [HAE-C1INH (Type 1 or Type 2)] confirmed by ONE of the following:
        (1) Diagnosis confirmed with measurements of C1-INH protein level, C1-INH function level, and C4 level to classify as Type 1 or Type 2 as identified by prescriber **or**
        (2) Diagnosis confirmed by mutation in the C1-INH gene altering protein synthesis and/or function as identified by prescriber **or**
     b) Diagnosis of normal C1INH (HAE-nl-C1INH) confirmed with levels within the normal range for C1-INH protein level, C1-INH function level, and C4 level as identified by prescriber **and**
        (1) ONE of the following:
            (a) Diagnosis is associated with a mutation in ONE of the following genes:
                (i) Coagulation factor FXII (mutation in F12)
                (ii) Plasminogen
                (iii) Angiopoietin-1
                (iv) Kininogen1
                (v) Heparan sulfate 3-O-sulfotransferase 6 gene
                (vi) Myoferlin gene **or**
  3. Diagnosis of HAE-unknown mutation (HAE-U) that has been confirmed by an HAE specialist (medical records required) **and**
  4. Patient's age is 18 years old or there is support for using the requested agent for the patient's age for the requested indication **and**
  5. Chart notes and/or prescriber do not provide documentation of concurrent use

of medications known to cause angioedema (i.e., ACE-Inhibitors, estrogens, angiotensin II receptor blockers) or prescriber has documented that the cause of angioedema is not related to use of these medications **and**

6. Prescriber is a specialist or has consulted with a specialist in the area of the patient's diagnosis (e.g., allergy and immunology, HAE specialist) **and**
7. Chart notes and/or prescriber do not provide documentation of concurrent use of other agents used to treat acute HAE (i.e., Berinert, Firazyr, icatibant, Kalbitor, Ruconest, Sajazir, Ekterly)

Approval Duration: 3 months

B. Hereditary angioedema due to C1INH deficiency [HAE-C1INH (Type 1 or Type 2)], prophylaxis as indicated by chart notes within past 90 days

1. Requested drug is Haegarda and will be used for long-term prophylaxis **and**
2. Diagnosis confirmed by ONE of the following:
    a) Diagnosis confirmed with measurements of C1-INH protein level, C1-INH function level, and C4 level to classify as Type 1 or Type 2 as identified by prescriber **or**
    b) Diagnosis confirmed by mutation in the C1-INH gene altering protein synthesis and/or function as identified by prescriber **and**
3. Patient has a history of at least three moderate to severe acute HAE attacks per month (e.g., airway swelling, severe abdominal pain, painful facial swelling), or one throat or upper respiratory life-threatening swelling per month **and**
4. Patient's age is within FDA labeling for the requested indication or there is support for using the requested agent for the patient's age for the requested indication **and**
5. Chart notes and/or prescriber do not provide documentation of concurrent use for medications known to cause angioedema (i.e., ACE-Inhibitors, estrogens, angiotensin II receptor blockers) or prescriber has documented that the cause of angioedema is not related to use of these medication **and**
6. Prescriber is an HAE specialist **and**
7. Chart notes and/or prescriber do not provide documentation of concurrent use of other agents used for prophylaxis of HAE attacks (i.e., Cinryze, Takhzyro, Orladeyo, Andembry)

Approval Duration: 12 months

II. Continued Therapy Criteria

A. Hereditary angioedema, acute treatment as indicated by chart notes within past 12 months

1. Requested drug is icatibant **and**

2. Patient has been previously approved for the requested agent through the plan's Prior Authorization process or meets the initial therapy criteria above **and**
3. Documented clinical benefit since starting the requested agent (i.e., decrease in duration of HAE attacks, quick onset of symptom relief, complete resolution of symptoms, or decrease in attack frequency/severity) **and**
4. Chart notes and/or prescriber do not provide documentation of concurrent use of other agents indicated for the treatment of acute HAE attacks (i.e., Berinert, Firazyr, Kalbitor, Ruconest, Sajazir, Ekterly) **and**
5. Prescriber is a specialist or has consulted with a specialist in the area of the patient's diagnosis (e.g., allergy and immunology, HAE specialist)

Approval Duration: 12 months

B. Hereditary angioedema, prophylaxis as indicated by chart notes within past 12 months

1. Requested drug is Haegarda **and**
2. Patient has been previously approved for the requested agent through the plan's Prior Authorization process or meets the initial therapy criteria above **and**
3. Documented clinical benefit since starting the requested agent by at least ONE of the following:
    a) A decrease in the frequency of HAE attacks from baseline (prior to treatment) **or**
    b) A decrease in HAE attack severity **or**
    c) A decrease in duration of HAE attacks **and**
4. Chart notes and/or prescriber do not provide documentation of concurrent use of other agents indicated for prophylaxis of HAE attacks (i.e., Cinryze, Takhzyro, Orladeyo, Andembry) **and**
5. Prescriber is an HAE specialist

Approval Duration: 12 months

Policy Owned by: Curative PBM team