

Hereditary Angioedema (HAE) Prior Authorization

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

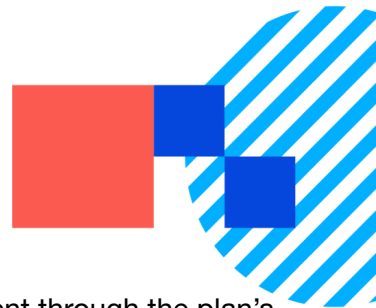
1. Requested drug is Haegarda and will be used for long-term prophylaxis **and**
2. Diagnosis confirmed by ONE of the following:
 - a) Diagnosis confirmed with measurements of C1-INH protein level, C1-INH function level, and C4 level to classify as Type 1 or Type 2 as identified by prescriber **or**
 - 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