

Hereditary Angioedema (HAE) Drug Products

DURB Approval Dates	07/2019; 10/2025
Commissioners' Approval Dates	2019; 2/2026

Protocol applies to FDA approved biosimilars and related indications and dosages

Addendum

The protocol was updated to include: Andembry, Dawnzera Ekterly, and Orladeyo.

Background

Hereditary angioedema (HAE) is a rare autosomal dominant genetic disorder. In patients with HAE a mutation to the C1-esterase-inhibitor (C1-INH) located on Chromosome 11q results in low levels of C1-INH activity and low levels of C4. C1-INH regulates activation of the complement and intrinsic coagulation (contact system pathway) and is a major inhibitor of plasma kallikrein. The kallikrein-kinin system is involved in the inflammatory and coagulation pathways. Kallikrein converts high molecular weight (HMW) kininogen to bradykinin. Bradykinin is a vasodilator that leads to swelling, inflammation and pain in patients with HAE.

Preferred Agents for Prophylaxis:

Haegarda (C1 inhibitor)

Takhzyro (lanadelumab)

Non-Preferred Agent for Prophylaxis:

Cinryze (C1inhibitor)

Andembry (garadacimab-gxii)

Dawnzera (donidalorsen)

Orladeyo (berotralstat)

Preferred Agents for Acute treatment:

Berinert (C1 inhibitor)

Icatibant

Sajazir (icatibant)

Non-Preferred Agents for Acute treatment:

Firazyr (icatibant)

Kalbitor (ecallantide)

Ruconest (C1 inhibitor)

Ekterly (sebetralstat)

General Criteria for Approval (must meet all of the following):

1. The patient is unable to take the preferred agents for the given diagnosis, due to a trial and inadequate treatment response or intolerance, or a contraindication.
Documentation is required for approval.
2. Patient is of the FDA-labeled or compendial approved age
3. A confirmed diagnosis of Hereditary Angioedema and documentation has been received
4. The medication requested is prescribed in accordance with a Food and Drug Administration (FDA) established indication and dosing regimen or in accordance with a

medically appropriate off-label indication and dosing according to American Hospital Formulary Service, Micromedex,

5. Clinical Pharmacology, Wolters Kluwer Lexi-Drugs (Lexicomp), national guidelines, or other peer-reviewed evidence
6. Patient does not have any contraindications to therapy
7. Patient's current weight is provided
8. Medication is prescribed by or in consultation with an allergist, immunologist, or a physician that specializes in the treatment of HAE or related disorders
9. Medications known to cause angioedema have been evaluated and discontinued when appropriate
10. The patient will not use the requested medication concurrently with another drug within the same treatment category (i.e. no more than one drug for prophylaxis or more than one drug for acute attacks)

Criteria for Continuation of Therapy:

1. Patient meets initial criteria for approval
2. Patient has documented improvement or does not have clinical worsening of HAE

Note: Kalbitor – Anaphylaxis reported post administration. Kalbitor should only be administered by a healthcare professional with appropriate medical support to manage anaphylaxis and HAE. The healthcare professional should be able to distinguish between hypersensitivity reactions and HAE related symptoms.

Approval Duration: 6 months

References:

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10. Dawnzera [package insert]. Ionis Pharmaceuticals Inc. Carlsbad, CA. 8/2025
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