BlueCross BlueShield
Federal Employee Program.
Federal Employee Program®
1310 G Street, N.W.
Washington, D.C. 20005
202.942.1000
Fax 202.942.1125

5.85.26

Section:	Prescription Drugs	**Effective Date:**	April 1, 2021
Subsection:	Hematological Agents	**Original Policy Date:**	July 14, 2017
Subject:	Haegarda	**Page:**	1 of 4

Last Review Date: March 12, 2021

Haegarda

Description

Haegarda (C1 esterase inhibitor [human])

Background
Haegarda is a C1-esterase inhibitor used for the routine prophylaxis against angioedema attacks with hereditary angioedema (HAE). HAE is caused by having insufficient amounts of a plasma protein called C1-esterase inhibitor. People with HAE can develop rapid swelling of the hands, feet, limbs, face, intestinal tract, or airway. These acute attacks of swelling can occur spontaneously, or can be triggered by stress, surgery, or infection. Swelling of the airway is potentially fatal without immediate treatment. Haegarda is intended to restore the level of functional C1-esterase inhibitor in a patient's plasma, thereby preventing the acute attack of swelling (1-4).

Regulatory Status
FDA-approved indication: is a plasma-derived concentrate of C1 Esterase Inhibitor (Human) (C1-INH) indicated for routine prophylaxis to prevent Hereditary Angioedema (HAE) attacks in patients 6 years of age and older (2).

Hypersensitivity reactions may occur. Epinephrine should be immediately available to treat any acute severe hypersensitivity reactions following discontinuation of administration (2).

Thrombotic events have been reported at the recommended dose of C1 Esterase Inhibitor (human) products, including Haegarda, following treatment of HAE. Monitor closely patients with known risk factors for thrombotic events (2).

Related policies
Berinert, Cinryze, Firazyr, Kalbitor, Orladeyo, Ruconest, Takhzyro

Policy

This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.

Haegarda may be considered **medically necessary** in patients 6 years of age or older for the routine prevention of hereditary angioedema (HAE) attacks and if the conditions indicated below are met.

Haegarda may be considered **investigational** in patients less than 6 years of age and for all other indications.

Prior-Approval Requirements

Age 6 years of age and older

Diagnosis

Patient must have **ALL** of the following:

1. Hereditary Angioedema (HAE)
    a. Routine prevention of angioedema attacks
    b. **NO** dual therapy with other agents for the prevention of hereditary angioedema attacks
    c. Inadequate treatment response or intolerance to a short-term course (5-days or less) of an androgen such as danazol, or a contraindication to one such as:
        i. Undiagnosed abnormal genital bleeding
        ii. Markedly impaired hepatic, renal, or cardiac function
        iii. Pregnancy (member is currently pregnant or may become pregnant)
        iv. Breast feeding
        v. Porphyria
        vi. Androgen-dependent tumor
        vii. Active thrombosis or history of thromboembolic disease
        viii. Prepubertal child

Prior – Approval *Renewal* Requirements

Age 6 years of age and older

Diagnosis

Patient must have **ALL** of the following:

1. Hereditary Angioedema (HAE)
    a. Routine prevention of angioedema attacks
    b. **NO** dual therapy with other agents for the prevention of hereditary angioedema attacks

Policy Guidelines

Pre - PA Allowance

None

Prior - Approval Limits

Duration 12 months

Prior – Approval *Renewal* Limits

Same as above

Rationale

Summary

Haegarda is a C1 esterase inhibitor indicated for routine prophylaxis against angioedema attacks in patients 6 years of age and older with Hereditary Angioedema (HAE). HAE symptoms include episodes of edema (swelling) in various body parts including the hands, feet, face, and airway. HAE is caused by mutations to C1-esterase-inhibitor (C1-INH). Serious arterial and venous thromboembolic (VTE) events have been reported at the recommended dose of plasma derived C1 esterase inhibitor products in patients with risk factors. The safety and efficacy of Haegarda in children less than 6 years of age has not been established. Persons who experience frequent and/or severe episodes may be candidates for prophylactic treatment (1-4).

Prior authorization is required to ensure the safe, clinically appropriate and cost-effective use of Haegarda while maintaining optimal therapeutic outcomes.

References

1. Zuraw BL. Clinical practice. Hereditary angioedema. N Engl J Med. Sep 4 2008;359(10):1027-36.2.
2. Haegarda. [package insert]. Kankakee, IL; CSL Behring LLC; September 2020.
3. Zuraw BL, Banerji A, Bernstein JA, et al. US Hereditary Angioedema Association Medical Advisory Board 2013 recommendations for the management of hereditary angioedema due to C1 inhibitor deficiency. J Allergy Clin Immunol: In Practice. 2013; 1(5): 458-467.
4. Wintenberger C, Boccon-Gibod I, Launay D, et al. Tranexamic acid as maintenance treatment for non-histaminergic angioedema: analysis of efficacy and safety in 37 patients. Clin Exp Immunol 2014; 178:112.

Policy History

Date	Action
July 2017	Addition to PA
September 2017	Annual review
December 2017	Annual editorial review and reference update Addition of inadequate treatment response, intolerance, or contraindication to a danazol or tranexamic acid per SME
March 2018	Annual review
September 2018	Changed wording of no dual therapy requirement
November 2018	Annual editorial review and reference update. Removal of requirement to try and fail tranexamic acid and reworded danazol or androgen trial requirement per SME
September 2019	Annual review
September 2020	Annual review and reference update
December 2020	Age requirement reduced from 12 and older to 6 and older
March 2021	Annual editorial review

Keywords

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on March 12, 2021 and is effective on April 1, 2021.