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). Hereditary angioedema, which 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 adolescent and adult patients (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).
Haegarda may be considered medically necessary in patients 12 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 12 years of age and for all other indications.

Prior-Approval Requirements

Age

12 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. Porphyrria
      vi. Androgen-dependent tumor
      vii. Active thrombosis or history of thromboembolic disease
      viii. Prepubertal child
Prior – Approval Renewal Requirements

Age
12 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

Duration
12 months

Rationale

Summary
Haegarda is a C1 esterase inhibitor indicated for routine prophylaxis against angioedema attacks in adolescent and adult patients 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 12 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

Policy History

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<tr>
<td>July 2017</td>
<td>Addition to PA</td>
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<td>September 2017</td>
<td>Annual review</td>
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<tr>
<td>December 2017</td>
<td>Annual editorial review and reference update. Addition of inadequate treatment response, intolerance, or contraindication to a danazol or tranexamic acid per SME</td>
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<tr>
<td>March 2018</td>
<td>Annual review</td>
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<tr>
<td>September 2018</td>
<td>Changed wording of no dual therapy requirement</td>
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<tr>
<td>November 2018</td>
<td>Annual editorial review and reference update. Removal of requirement to try and fail tranexamic acid and reworded danazol or androgen trial requirement per SME</td>
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Keywords

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on September 13, 2019 and is effective on October 1, 2019.