**Firazyr (icatibant)**

**Last Review Date: June 2, 2017**
**Number: MG.MM.PH.34**

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**Definition**

Hereditary angioedema is an autosomal dominant condition marked by unpredictable and recurrent potentially fatal angioedema attacks; treatment options include injectable drugs for prophylaxis or for acute attacks.

**Initial Guideline**

Firazyr (icatibant) is considered medically necessary for the treatment of acute attacks of hereditary angioedema and coverage will be provided for 12 weeks if the member meets all of the following criteria:

1. > 18 years of age

2. Must be prescribed by, or in consultation with, a specialist in: Allergy, immunology, hematology, pulmonology, or medical genetics

3. Confirmation that member is avoiding the following possible triggers for HAE attacks:
   - Helicobacter pylori infections (confirmed by lab test)
   - Estrogen-containing oral contraceptive agents OR hormone replacement therapy
   - Antihypertensive agents containing ACE inhibitors

4. Member has a history of moderate to severe cutaneous or abdominal attacks OR mild to severe airway swelling attacks of HAE (i.e. debilitating cutaneous/gastrointestinal symptoms or laryngeal/pharyngeal/tongue swelling)

5. Member has one of the following clinical presentations (table below) consistent with HAE subtype:
**HAE I**
- Low C1 inhibitor- (C1-INH) antigenic level (C1-INH antigenic level below the lower limit of normal as defined by the laboratory performing the test); AND
- Low C4 level (C4 below the lower limit of normal as defined by the laboratory performing the test); AND
- Low C1-INH functional level (C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test); AND
- Member has a family history of HAE; OR
- Normal C1q level

**HAE II**
- Normal to elevated C1-INH antigenic level; AND
- Low C4 level (C4 below the lower limit of normal as defined by the laboratory performing the test); AND
- Low C1-INH functional level (C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test)

**HAE III**
- Normal C1-INH antigenic level; AND
- Normal C4 level; AND
- Normal C1-INH functional level; AND
- Member has a known HAE-causing C1-INH mutation (i.e., mutation of coagulation factor XII gene); OR
- Member has a family history of HAE

**Renewal Guideline**

Coverage will be provided for 12 weeks when **all** of the following criteria are met:

1. Member continues to meet the criteria in the initial guideline
2. Significant improvement in severity and duration of attacks have been achieved and sustained
3. Absence of unacceptable toxicity from the drug (e.g., hypersensitivity reactions, thrombotic events, laryngeal attacks)
4. The cumulative amount of medication(s) the member has on-hand, indicated for the acute treatment of HAE, will be taken into account when authorizing. The authorization will provide a sufficient quantity in order for the member to have a cumulative amount of HAE medication(s) on-hand in order to treat up to 4 acute attacks per 4 weeks for the duration of the authorization.

**Limitations/Exclusions**

Use of Firazyr (icatibant) is considered experimental or investigational for all other uses.

**Applicable Procedure Codes**

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<thead>
<tr>
<th>Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>J1744</td>
<td>Injection, icatibant, 1mg</td>
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**Applicable Diagnosis Codes**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>D84.1</td>
<td>Defects in the complement system</td>
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References


7. Specialty-matched clinical peer review.