Definition
FIRAZYR is a bradykinin B2 receptor antagonist indicated for treatment of acute attacks of hereditary angioedema (HAE) in adults 18 years of age and older.

Dosing
Max dose (per dose and over time):
• 30 billable units per dose up to 3 times in a 24-hour period once per week

Guideline
Firazyr (icatibant) is considered medically necessary for the treatment of acute attacks of hereditary angioedema when the following criteria are met:

• Patient is 18 years of age or older; AND
• Firazyr is prescribed by, or in consultation with, a specialist in: allergy, immunology, hematology, pulmonology, or medical genetics; AND
• Confirmation that member is avoiding medications known to cause angioedema (e.g., ACE inhibitors, oral contraceptives, hormone replacement therapy); AND
• 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); AND
• Member has one of the following clinical presentations (table below) consistent with HAE subtype:

HAE I (C1-inhibitor deficiency)
- 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 (C1-inhibitor dysfunction)
- 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 with normal C1NH (HAE III)
- Normal C1-INH antigenic level; AND
- Normal C4 level; AND
- Normal C1-INH functional level; AND
  - Member has a known HAE-causing mutation (e.g., mutation of coagulation factor XII gene [F12 mutation], mutation in the angiopoietin-1 gene, mutation in the plasminogen gene); OR
  - Member has a family history of HAE

Coverage for Firazyr (icatibant) may be renewed when the subsequent criteria are met:
- Member continues to meet the criteria in the initial guideline; AND
- Significant improvement in severity and duration of attacks have been achieved and sustained; AND
- Absence of unacceptable toxicity from the drug (e.g., hypersensitivity reactions, thrombotic events, laryngeal attacks); AND

### Limitations/Exclusions
- Approval will be granted for 6 months and may be renewed
- 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.
- Use of Firazyr (icatibant) is considered experimental or investigational for all other uses.

### Revision History
7/25/2018 – Increased coverage duration to 6 months, removed confirmation patient is negative for helicobacter pylori infection, updated table describing clinical presentations consistent with HAE

### Applicable Procedure Codes
Applicable Diagnosis Codes

<table>
<thead>
<tr>
<th>J1744</th>
<th>Injection, icatibant, 1mg</th>
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<tr>
<td>D84.1</td>
<td>Defects in the complement system</td>
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References


7. Specialty-matched clinical peer review.