Reference number(s)
2192-A

STANDARD MEDICARE PART B MANAGEMENT

FIRAZYR (icatibant)
Sajazir (icatibant)
icatibant

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

A. FDA-Approved Indication
Treatment of acute attacks of hereditary angioedema (HAE) in adults 18 years of age and older

B. Compendial Use
Treatment of angiotensin-converting enzyme (ACE) inhibitor-induced angioedema

All other indications will be assessed on an individual basis. Submissions for indications other than those enumerated in this policy should be accompanied by supporting evidence from Medicare approved compendia.

II. DOCUMENTATION

The following documentation must be available, upon request, for all submissions:
A. For initial authorization:
1. C4 levels and C1 inhibitor functional and antigenic protein levels
2. F12, angiopoietin-1, plasminogen, kininogen-1 (KNG1), heparan sulfate-glucosamine 3-O-sulfotransferase 6 (HS3ST6), or myoferlin (MYOF) gene mutation testing, if applicable
3. Chart notes confirming family history of angioedema, if applicable

B. For continuation of therapy, chart notes demonstrating a reduction in severity and/or duration of attacks

III. CRITERIA FOR INITIAL APPROVAL

A. Hereditary angioedema (HAE)
Authorization of 6 months may be granted for treatment of acute HAE attacks when either of the following criteria is met:
1. Member has C1 inhibitor deficiency or dysfunction as confirmed by laboratory testing and meets both of the following criteria:
   a. Member has a C4 level below the lower limit of normal as defined by the laboratory performing the test, and
   b. Member meets one of the following criteria:

i. C1 inhibitor (C1-INH) antigenic level below the lower limit of normal as defined by the laboratory performing the test, or
ii. Normal C1-INH antigenic level and a low C1-INH functional level (functional C1-INH less than 50% or C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test).

2. Member has normal C1 inhibitor as confirmed by laboratory testing and meets one of the following criteria:
   a. Member has an F12, angiopoietin-1, plasminogen, kininogen-1 (KNG1), heparan sulfate-glucosamine 3-O-sulfotransferase 6 (HS3ST6), or myoferlin (MYOF) gene mutation as confirmed by genetic testing.
   b. Member has a documented family history of angioedema and the angioedema was refractory to a trial of high-dose antihistamine therapy (i.e., cetirizine at 40 mg per day or the equivalent) for at least one month.

B. ACE inhibitor-induced angioedema
Authorization of 3 days may be granted for acute management of ACE inhibitor-induced angioedema.

IV. CONTINUATION OF THERAPY

A. Hereditary angioedema
All members (including new members) requesting authorization for continuation of therapy must be currently receiving therapy with the requested agent.

Authorization for 12 months may be granted for treatment of acute HAE attacks when all of the following criteria are met:
1. The member is currently receiving therapy with the requested drug.
2. The requested drug is being used to treat an indication enumerated in Section III.
3. The member is receiving benefit from therapy. Benefit is defined as a reduction in severity and/or duration of attacks.

B. ACE inhibitor-induced angioedema
All members (including new members) requesting reauthorization for continuation of therapy must meet all initial authorization criteria.

V. REFERENCES

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15. Veronez CL, Csuka D, Sheik FR, et al. The expanding spectrum of mutations in hereditary angioedema. *J Allergy Clin Immunol Pract.* 2021;S2213-2198(21)00312-3.
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