Orladeyo (berotralstat)

Summary

Orladeyo (berotralstat) is FDA-approved for prophylaxis to prevent attacks of hereditary angioedema (HAE) in adults and pediatrics 12 years of age and older. HAE is a rare disease that causes swelling, pain and inflammation in various areas of the body including the face, hands, feet, throat, stomach, and bowels. HAE can be caused by a reduced amount or decreased function of C1 esterase inhibitor protein, a regulator of inflammatory pathways.

The recommended dosage for Orladeyo (berotralstat) is 150 mg taken orally once daily. It is available as 150 mg and 110 mg capsules because dosage may need to be adjusted for certain individuals (e.g., with moderate or severe hepatic impairment or persistent gastrointestinal reactions). Alternatives for Orladeyo (berotralstat) include products such as Takhzyro (lanadelumab-flyo), Haegarda (C1 inhibitor, concentrate from human plasma), Cinryze (C1 inhibitor, concentrate from human plasma), or Dawnzera (donidalorsen).

Definitions

"C1 inhibitor" refers to an enzyme that functions as a major anti-inflammatory protein in the body. People who have HAE have low levels of C1 inhibitor in their body.

"C4" or "Complement 4" refers to an enzyme that is involved in the inflammatory response. Alterations in C1 inhibitor can result in low levels of C4 in people who have HAE.

"Hereditary angioedema (HAE)" refers to a rare, inherited disease that causes swelling, pain and inflammation in various areas of the body including the face, hands, feet, throat, stomach, and bowels.

Medical Necessity Criteria for Initial Authorization

The Plan considers Orladeyo (berotralstat) medically necessary when ALL of the following criteria are met:

1. Prescribed by or in consultation with an allergist, hematologist, immunologist, or other specialist experienced in the diagnosis and management of hereditary angioedema (HAE)*; AND*
2. The member is 12 years of age or older*; AND*
3. Is being used for prevention of attacks of hereditary angioedema (HAE), confirmed by ONE (1) of the following:
    a. Clinically appropriate low levels (as defined by the laboratory reference values) of BOTH:
        i. Low complement C4 (either at baseline or during an attack)*; and*
        ii. Low C1 esterase inhibitor (C1-INH) antigenic protein level (Type 1 HAE) OR functional level (Type 2 HAE)*; or*
    b. Documented normal or near normal C4, C1-INH antigen, and C1-INH function AND ONE of the following:
        i. Demonstration of a mutation (e.g., in the factor XII, plasminogen, angiopoietin-1, SERPING1, kininogen gene, Myoferlin [MYOF], or Heparan sulfate glucosamine 3-O-sulfotransferase 6 [HS3ST5]) associated with HAE*; or*
        ii. A positive family history of recurrent angioedema or C1-INH deficiency and documented lack of efficacy of high-dose antihistamine therapy (e.g., cetirizine at 40 mg/day or the equivalent)*; AND*
4. The member has a documented history of moderate or severe attacks of hereditary angioedema (e.g., airway swelling, severe abdominal pain, facial swelling, nausea and vomiting, painful facial distortion)*; AND*
5. Dosage is within the recommended limit of 150 mg (taken orally) once daily*; AND*
6. Chart documentation and supporting laboratory test results are provided for review to substantiate the above listed requirements.

If the above prior authorization criteria are met, Orladeyo (berotralstat) will be approved for up to 12-months.

Medical Necessity Criteria for Reauthorization

Reauthorization for up to 12 months will be granted if BOTH of the following are met:

1. The member still meets all applicable initial criteria*; AND*
2. Chart documentation shows the member has experienced a positive clinical response to therapy as evidenced by ONE (1) of the following compared to baseline (prior to starting prophylaxis therapy):
    a. The member has experienced a significant reduction in frequency of attacks (e.g., ≥ 50%) since starting prophylactic treatment; *or*
    b. The member has reduced the use of medications to treat acute attacks since starting prophylactic treatment. .

Experimental or Investigational / Not Medically Necessary

Orladeyo (berotralstat) for any other indication is considered not medically necessary by the Plan, as it is deemed to be experimental, investigational, or unproven.

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