Keveyis

Description

Keveyis (dichlorphenamide)

Background
Keveyis is an oral carbonic anhydrase inhibitor indicated for the treatment of periodic paralysis. Periodic paralyses are a group of rare hereditary disorders that cause episodes of muscle weakness or paralysis. Types of periodic paralyses are differentiated by criteria including underlying genetic mutations and changes in blood-potassium during attack. Hypokalemic and hyperkalemic are two common types of periodic paralyses (1).

Regulatory Status
FDA-approved indications: Keveyis is an oral carbonic anhydrase inhibitor indicated for the treatment of primary hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, and related variants (2).

Keveyis includes a contraindication for hepatic insufficiency. Keveyis may aggravate hepatic encephalopathy. Keveyis also includes a contraindication for severe pulmonary disease. Keveyis can cause hyperchloremic non-anion gap metabolic acidosis. Patients with severe pulmonary disease may be unable to compensate for the metabolic acidosis caused by Keveyis. Concomitant use of Keveyis with other drugs that cause metabolic acidosis may increase the severity of metabolic acidosis. Baseline and periodic measurement of serum bicarbonate during Keveyis treatment are recommended. If metabolic acidosis develops or persists, consider reducing the dose or discontinuing Keveyis (2).
The use of Keveyis is contraindicated with concomitant use of high-dose aspirin. Anorexia, tachypnea, lethargy, and coma have been reported with co-administration of high-dose aspirin and Keveyis. Keveyis should be used with caution in patients receiving low-dose aspirin (2).

Keveyis increases potassium excretion and can cause hypokalemia. Baseline and periodic measurement of serum potassium are recommended. If hypokalemia develops or persists, consider reducing the dose or discontinuing Keveyis (2).

The safety and efficacy of Keveyis in pediatric patients 18 years or less have not been established (2).

Related policies

Keveyis may be considered medically necessary in patients 18 years of age or older for the treatment of primary hyperkalemic or hypokalemic periodic paralysis and related variants with baseline and periodic monitoring of serum potassium and bicarbonate levels; diagnosis confirmed by one of the following: genetic testing, provocative testing, electromyography, or muscle biopsy; documentation that lifestyle modifications, dietary restrictions and exercise restrictions have been maximally challenged; inadequate treatment response, intolerance, or contraindication to acetazolamide; no signs of hepatic impairment; no severe pulmonary disease; and no use of high dose aspirin.

Keveyis is considered investigational in patients who are less than 18 years of age and for all other indications.

Prior-Approval Requirements

Age

18 years of age or older

Diagnoses

Patient must have ONE of the following:

1. Primary hyperkalemic periodic paralysis and related variants
2. Primary hypokalemic periodic paralysis and related variants

AND ALL of the following:

1. Baseline and periodic monitoring of serum potassium and bicarbonate levels
2. Diagnosis confirmed by ONE of the following:
   a. Genetic testing
   b. Provocative testing
   c. Electromyography
   d. Muscle biopsy
3. Documentation that lifestyle modifications, dietary restrictions and exercise restrictions have been maximally challenged
4. Inadequate treatment response, intolerance, or contraindication to acetazolamide

AND NONE of the following:
1. Signs of hepatic impairment
2. Severe pulmonary disease
3. Use of high-dose aspirin

Prior – Approval Renewal Requirements

Age 18 years of age or older

Diagnoses

Patient must have ONE of the following:

1. Primary hyperkalemic periodic paralysis and related variants
2. Primary hypokalemic periodic paralysis and related variants

AND the following:

1. Documentation that the patient has had a reduction in the number of paralytic attacks

AND NONE of the following:
1. Signs of hepatic impairment
2. Severe pulmonary disease
3. Use of high-dose aspirin
Policy Guidelines

Pre - PA Allowance

None

Prior - Approval Limits

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<th>Quantity</th>
<th>360 tablets per 90 days</th>
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<tr>
<td>Duration</td>
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Prior – Approval Renewal Limits

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Rationale

Summary
Keveyis is an oral carbonic anhydrase inhibitor indicated for the treatment of primary hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, and related variants. Keveyis has an unknown mechanism of therapeutic effect on patients with periodic paralysis. Keveyis can cause metabolic acidosis and use is contraindicated in patients with severe pulmonary disease. Keveyis may aggravate hepatic encephalopathy and use is contraindicated in patients with hepatic impairment. Co-administration of Keveyis with high-dose aspirin is contraindicated due to the risk of coma. Monitoring of potassium and bicarbonate levels is required at baseline and periodically throughout treatment with Keveyis. The safety and efficacy of Keveyis in pediatric patients 18 years or less have not been established (1-2).

Prior authorization is required to ensure the safe, clinically appropriate and cost-effective use of Keveyis while maintaining optimal therapeutic outcomes.

References
**Section:** Prescription Drugs  
**Effective Date:** October 1, 2019  
**Subsection:** Cardiovascular Agents  
**Original Policy Date:** November 6, 2015  
**Subject:** Keveyis  
**Page:** 5 of 5

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<thead>
<tr>
<th>Date</th>
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<tr>
<td>November 2015</td>
<td>Addition to PA</td>
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| December 2015| Annual editorial review  
Addition of lifestyle, dietary and exercise requirements and trial of acetazolamide per PMPC |
| December 2016| Annual editorial review  
Addition of age to renewal requirements  
Policy number change from 5.16.09 to 5.40.09 |
| September 2017| Annual review and reference update                                     |
| September 2018| Annual review and reference update                                     |
| September 2019| Annual review                                                          |

**Keywords**

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