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# 5.40.009

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| **Section:** | Prescription Drugs | **Effective Date:** | July 1, 2023 |
| **Subsection:** | Cardiovascular Agents | **Original Policy Date:** | November 6, 2015 |
| **Subject:** | Keveyis | **Page:** | 1 of 5 |

***Last Review Date:*** June 15, 2023

# Keveyis

## Description

### Keveyis (dichlorphenamide)

**Background**
Keveyis (dichlorphenamide) 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**

## Policy

*This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.*

Keveyis may be considered **medically necessary** if the conditions indicated below are met.

Keveyis may be considered **investigational** 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

**Quantity** 360 tablets per 90 days

**Duration** 3 months

## Prior – Approval *Renewal* Limits

**Quantity** 360 tablets per 90 days

**Duration** 12 months

## Rationale

**Summary**
Keveyis (dichlorphenamide) 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**
1. National Institute of Neurological Disorders and Stroke. NINDS Familial Periodic Paralyses Information Page. https://www.ninds.nih.gov/Disorders/All-Disorders/Familial-Periodic-Paralyses-Information-Page. Published May 24, 2017.
2. Keveyis [package insert]. Hawthorne, NY. Taro Pharmaceuticals USA, Inc.; November 2019.

## Policy History

| Date | Action |
|---|---|
| November 2015 | Addition to PA |
| December 2015 | Annual editorial review<br>Addition of lifestyle, dietary and exercise requirements and trial of acetazolamide per PMPC |

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| December 2016 | Annual editorial review<br>Addition of age to renewal requirements<br>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 |
| September 2020 | Annual review and reference update |
| June 2021 | Annual review |
| June 2022 | Annual review |
| June 2023 | Annual review. Changed policy number to 5.40.009 |

## Keywords

**This policy was approved by the FEP® Pharmacy and Medical Policy Committee on June 15, 2023 and is effective on July 1, 2023.**