



## **KEVEYIS** (dichlorphenamide)

### **RATIONALE FOR INCLUSION IN PA PROGRAM**

#### **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).

#### **Summary**

Keveyis (dichlorphenamide) is an oral carbonic anhydrase inhibitor indicated for the treatment of



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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 Paralysis Information Page. <https://www.ninds.nih.gov/Disorders/All-Disorders/Familial-Periodic-Paralysis-Information-Page>. Published May 24, 2017.
2. Keveyis [package insert]. Hawthorne, NY. Taro Pharmaceuticals USA, Inc. January 2024.