Pharmacy Policy Bulletin: J-0764 Dichlorphenamide Products – Commercial and Healthcare Reform	
Number: J-0764	**Category:** Prior Authorization
Line(s) of Business: ☒ Commercial ☒ Healthcare Reform ☐ Medicare	**Benefit(s):** **Commercial:** **Prior Authorization** 1. Miscellaneous Specialty Drugs Oral = Yes w/ Prior Authorization **Healthcare Reform:** Not Applicable
Region(s): ☒ All ☐ Delaware ☐ New York ☐ Pennsylvania ☐ West Virginia	**Additional Restriction(s):** None
Version: J-0764-010	**Original Date:** 09/02/2015
Effective Date: 10/08/2025	**Review Date:** 09/17/2025

Drugs Product(s):	• Keveyis (dichlorphenamide) • Ormalvi (dichlorphenamide)
FDA-Approved Indication(s):	• Treatment of primary hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, and related variants.

Background:	• Dichlorphenamide is an oral carbonic anhydrase inhibitor. However, the precise mechanism by which it exerts its therapeutic effects in patients with periodic paralysis is unknown.
	• Ormalvi is a generic formulation of dichlorphenamide.
	• Periodic paralysis (PP) is a muscle disease that causes episodic muscle weakness or temporary paralysis. It is a rare, autosomal-dominant genetic neuromuscular disorder caused by mutations in skeletal muscle sodium, calcium, and potassium channel genes. The most common types of PP are hypokalemic periodic paralysis when episodes occur in association with low potassium blood levels and hyperkalemic periodic paralysis when episodes can be induced by elevated potassium.
	• Primary periodic paralysis is very rare with only 4,000 to 5,000 diagnosed individuals in the U.S.
	• Primary periodic paralyses include:
	  ○ Hypokalemic paralysis (HypoPP)
	  ○ Hyperkalemic paralysis (HyperPP)
	  ○ Anderson-Tawil syndrome
	• There are also closely related diseases whose features overlap with HypoPP and HyperPP, including paramyotonia congenita (PMC) and normokalemic PP.
	• ICD-10 Code Information:
	  ○ ICD-10: G72.3 “Periodic paralysis” member has a specific diagnosis of hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, or related variants.
	  ○ ICD-10: G71.19 “Other specified myotonic disorders” may apply to Keveyis; however, the prescriber must confirm that the member has a

	specific diagnosis of hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, or related variants. • Prescribing Considerations: ○ Dichlorphenamide is not indicated for treating glaucoma. The chemical entity dichlorphenamide was initially approved by the FDA in 1958 under the brand name Daranide for the treatment of glaucoma but was discontinued in June 2002. ○ Dichlorphenamide dose should not exceed the recommended maximum dose of 200 mg per day. ○ Dichlorphenamide is contraindicated with concomitant use of high dose aspirin, in patients with severe pulmonary obstruction, in patients with hepatic insufficiency, and in the presence of a hypersensitivity or allergy to sulfonamides. ○ Dichlorphenamide should be discontinued at the first appearance of skin rash or any immune-mediated or other life-threatening adverse reaction. ○ Primary hyperkalemic period paralysis, primary hypokalemic periodic paralysis, and related variants are a heterogeneous group of conditions for which the response to Keveyis may vary. Prescribers should evaluate the patient's response after 2 months of treatment to decide whether dichlorphenamide should be continued.

Approval Criteria

I. Initial Authorization

When a benefit, coverage of Keveyis (dichlorphenamide) or Ormalvi may be approved when all of the following criteria are met **(A. through D.):**

A. The member is 18 years of age or older.

B. The member has one (1) of the following diagnoses (no ICD-10 code) **(1. or 2.):**

1. Primary hyperkalemic periodic paralysis and related variants

2. Primary hypokalemic periodic paralysis and related variants

C. The prescriber attests to the baseline number of muscle weakness attacks per week.

D. If the request is for brand Keveyis or Ormalvi, the member has experienced therapeutic failure or intolerance to generic dichlorphenamide.

II. Reauthorization

When a benefit, reauthorization or Keveyis (dichlorphenamide) or Ormalvi may be approved when all of the following criteria are met **(A. and B.)**:

A. The prescriber attests that the member has experienced a decrease from baseline in the number of muscle weakness attacks per week.

B. If the request is for brand Keveyis or Ormalvi, the member has experienced therapeutic failure or intolerance to generic dichlorphenamide.

III. An exception to some or all of the criteria above may be granted for select members and/or circumstances based on state and/or federal regulations.

Limitations of Coverage

I. Coverage of drug(s) addressed in this policy for disease states outside of the FDA-approved indications should be denied based on the lack of clinical data to support effectiveness and safety in other conditions unless otherwise noted in the approval criteria.

II. For Commercial or HCR members with a closed formulary, a non-formulary product will only be approved if the member meets the criteria for a formulary exception in addition to the criteria outlined within this policy.

Authorization Duration

Initial Authorization

- Commercial and HCR Plans: If approved, up to a 2 month authorization may be granted.

Reauthorization

- Commercial and HCR Plans: If approved, up to a 12 month authorization may be granted.

Automatic Approval Criteria

None.

References:

1. Keveyis [package insert]. Trevose, PA: Strongbridge US Inc.; November 2019.
2. Ormalvi [package insert]. Cambridge, CB3 0FA, United Kingdom: Cycle Pharmaceuticals Ltd.; February 2024.
3. Periodic Paralysis Association: Primary Hypokalemic Periodic Paralysis, Primary Hypokalemic Periodic Paralysis. Available at: https://periodicparalysis.org/what-is-periodic-paralysis-2/primary-hypokalemic-periodic-paralysis/. Accessed July 7, 2025.
4. NIH-National Institute of Neurological Disorders and Stroke: NINDS Familial Periodic Paralysis Information Page. Available at: https://www.ninds.nih.gov/Disorders/All-Disorders/Familial-Periodic-Paralyses-Information-Page. Accessed July 7, 2025.
5. Statland, JM, Fontaine, B, Hanna, MG, et al. Review of the Diagnosis and Treatment of Periodic Paralysis. *Muscle Nerve* 57: 522-530, 2018.