

## Pharmacy Policy Bulletin

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**Title:** Dichlorphenamide (Keveyis®)

**Policy #:** Rx.01.176

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***Application of pharmacy policy is determined by benefits and contracts. Benefits may vary based on product line, group, or contract. Some medications may be subject to precertification, age, quantity, or formulary restrictions (ie limits on non-preferred drugs). Individual member benefits must be verified.***

***This pharmacy policy document describes the status of pharmaceutical information and/or technology at the time the document was developed. Since that time, new information relating to drug efficacy, interactions, contraindications, dosage, administration routes, safety, or FDA approval may have changed. This Pharmacy Policy will be regularly updated as scientific and medical literature becomes available. This information may include new FDA-approved indications, withdrawals, or other FDA alerts. This type of information is relevant not only when considering whether this policy should be updated, but also when applying it to current requests for coverage.***

***Members are advised to use participating pharmacies in order to receive the highest level of benefits.***

### **Intent:**

The intent of this policy is to communicate the medical necessity criteria for **dichlorphenamide (Keveyis®)** as provided under the member's prescription drug benefit.

### **Description:**

Periodic paralyses are a group of rare, inherited neuromuscular disorders, resulting from mutations in the genes that regulate muscle ion channels. These disorders are characterized by episodes of painless muscle weakness where the affected muscles become weak and unable to contract.

The two most common types of periodic paralyses are hypo- and hyper-kalemic. Hypokalemic periodic paralysis is characterized by a fall in potassium levels in the blood. In individuals with this mutation attacks often begin in adolescence and are triggered by strenuous exercise, high carbohydrate meals, or by injection of insulin, glucose, or epinephrine. Weakness may be mild and limited to certain muscle groups, or more severe and affect the arms and legs. Attacks may last for a few hours or persist for several days. Some patients may develop chronic muscle weakness later in life. Hyperkalemic periodic paralysis is characterized by a rise in potassium levels in the blood. Attacks often begin in infancy or early childhood and are precipitated by rest after exercise, fasting, cold exposure, or ingestion of small amounts of potassium. Attacks are usually shorter, more frequent, and less severe than the hypokalemic form. Muscle spasms are common. Weakness in an attack can be focal, affecting only one limb, but generalized weakness with hypotonia is more common.

Dichlorphenamide is an oral carbonic anhydrase inhibitor indicated for the treatment of primary hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, and related variants. The precise mechanism by which dichlorphenamide exerts its therapeutic effects in patients with periodic paralysis is unknown.

### **Policy:**

**INITIAL CRITERIA:** Dichlorphenamide (Keveyis®) is approved when ALL of the following are met:

1. Treatment of primary hyperkalemic periodic paralysis, primary hypokalemic paralysis, and related variants; and
2. Prescribed by or in consultation with a neurologist; and
3. No documentation of ANY of the following:
  - a. Concomitant use of high dose aspirin; or
  - b. Severe pulmonary disease; or
  - c. Hepatic insufficiency; and
  - d. Member is 18 years of age or older

Initial authorization duration: 3 months

**CONTINUATION CRITERIA:** Dichlorphenamide (Keveyis®) is reapproved when there is documentation of positive clinical response to therapy.

Continuation authorization duration: 2 years

**Black Box Warning as shown in the drug Prescribing Information:**

N/A

**Guidelines:**

Refer to the specific manufacturer's prescribing information for administration and dosage details and any applicable Black Box warnings.

**BENEFIT APPLICATION**

Subject to the terms and conditions of the applicable benefit contract, the applicable drug(s) identified in this policy is (are) covered under the prescription drug benefits of the Company's products when the medical necessity criteria listed in this pharmacy policy are met. Any services that are experimental/investigational or cosmetic are benefit contract exclusions for all products of the Company.

**References:**

Dichlorphenamide. Micromedex 2.0. Truven Health Analytics, Inc. Greenwood Village, CO. Available at: <http://www.micromedexsolutions.com/>. Accessed July 13, 2022.

Gutmann L, Conwit R. Hyperkalemic periodic paralysis. UpToDate. June 2014. Available at: <https://www.uptodate.com/contents/hyperkalemic-periodic-paralysis>. Accessed July 13, 2022.

Keveyis (dichlorphenamide) [package insert]. Hawthorne NY. Taro Pharmaceuticals USA, Inc. November 2019. Available at: [https://www.accessdata.fda.gov/drugsatfda\\_docs/label/2015/011366s030lbl.pdf](https://www.accessdata.fda.gov/drugsatfda_docs/label/2015/011366s030lbl.pdf). Accessed July 13, 2022

National Institute of Neurological Disorders and Stroke (NINDS). NINDS Familial Periodic Paralysis Information Page. Available at: <https://www.ninds.nih.gov/Disorders/All-Disorders/Familial-Periodic-Paralysis-Information-page>. Last updated March 27, 2019. Accessed July 13, 2022.

Gutmann L, Conwit R. Hyperkalemic periodic paralysis. UpToDate website. Last updated March 8, 2019. Available at: <http://www.uptodate.com/>. Accessed July 13, 2022.

**Applicable Drugs:**

Inclusion of a drug in this table does not imply coverage. Eligibility, benefits, limitations, exclusions, precertification/referral requirements, provider contracts, and Company policies apply.

Brand Name	Generic Name
Keveyis®	Dichlorphenamide

**Cross References:**

Off-Label Use Rx.01.33

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<b>Policy Version Number:</b>	8.00
<b>P&amp;T Approval Date:</b>	June 09, 2022
<b>Policy Effective Date:</b>	October 01, 2022
<b>Next Required Review Date:</b>	June 09, 2023

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The Policy Bulletins on this web site were developed to assist the Company in administering the provisions of the respective benefit programs, and do not constitute a contract. If you have coverage through the Company, please refer to your specific benefit program for the terms, conditions, limitations and exclusions of your coverage. Company does not provide health care services, medical advice or treatment, or guarantee the outcome or results of any medical

services/treatments. The facility and professional providers are responsible for providing medical advice and treatment. Facility and professional providers are independent contractors and are not employees or agents of the Company. If you have a specific medical condition, please consult with your doctor. The Company reserves the right at any time to change or update its Policy Bulletins.

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