



Policy Type: PA/SP

Pharmacy Coverage Policy: EOCCO121

Description

Dichlorphenamide (Keveyis) is a carbonic anhydrase inhibitor; however, the mechanism by which dichlorphenamide (Keveyis) exerts its therapeutic effects in patients with periodic paralysis is unknown.

Length of Authorization

- Initial: Two months
- Renewal: 12 months

Quantity Limits

| Product Name | Dosage Form | Indication | Quantity Limit |
|----------------------------|---------------|----------------------------|---------------------|
| dichlorphenamide (Keveyis) | 50 mg tablets | Primary periodic paralysis | 120 tablets/30 days |

Initial Evaluation

- I. Dichlorphenamide (Keveyis) may be considered medically necessary when the following criteria below are met:
 - A. Medication is prescribed by, or in consultation with, a neurologist or provider with experience in primary periodic paralysis (e.g. physiatrist); **AND**
 - B. A diagnosis of **periodic paralysis** when the following are met:
 1. Treatment with acetazolamide has been ineffective, contraindicated, or not tolerated.
- II. Dichlorphenamide (Keveyis) is considered investigational when used for all other conditions.

Renewal Evaluation

- I. Member has received a previous prior authorization approval for this agent through this health plan; **AND**
- II. Member is not continuing therapy based off being established on therapy through samples, manufacturer coupons, or otherwise. Initial policy criteria must be met for the member to qualify for renewal evaluation through this health plan; **AND**
- III. Member has exhibited improvement or stability of disease symptoms (e.g. reduced frequency or severity of paralytic attacks)

Supporting Evidence

- I. Periodic paralysis is a rare neuromuscular disorder related to a defect in muscle ion channels. It is classified as hypokalemic when episodes occur with low potassium levels and hyperkalemic when occurring with high. It is characterized by episodes of painless muscle paralysis, which may be precipitated by heavy exercise, fasting, or high-carbohydrate meals. Attacks may last minutes, hours, or days causing increased morbidity and impaired quality of life. Primary periodic paralyses include hypokalemic paralysis (HypoPP), hyperkalemic paralysis (HyperPP), and Andersen-Tawil syndrome. To prevent attacks, various methods are used including dietary modification, avoidance of triggers, potassium supplementation, and using carbonic anhydrase inhibitors.
- II. Keveyis is indicated for the treatment of primary hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, and related variants.
- III. Carbonic anhydrase inhibitors, particularly acetazolamide and dichlorphenamide, have been used for almost 50 years as empiric treatment for both HypoPP and HyperPP. There are no comparative studies between acetazolamide and dichlorphenamide to suggest greater safety or efficacy in one agent over another.
- IV. Per the package insert: Primary hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, and related variants are a heterogeneous group of conditions for which the response to KEVEYIS may vary. Therefore, prescribers should evaluate patient response to KEVEYIS after 2 months of treatment to determine whether KEVEYIS should be continued.
- V. Withdrawal from the study due to the acute and severe worsening of symptoms, for example, an increase in attack frequency or severity, was also assessed as an endpoint in clinical studies. Acute, intolerable worsening of condition was observed in 2/42 patients on KEVEYIS.

Investigational or Not Medically Necessary Uses

- I. Dichlorphenamide (Keveyis) has not been sufficiently evaluated outside of primary periodic paralysis.

References

1. Keveyis [Prescribing Information]. Hawthorne, NY: Taro Pharmaceuticals; November 2019.
2. UpToDate, Inc. Hypokalemic periodic paralysis. UpToDate [database online]. Waltham, MA. Last updated October 25, 2018 Available at: <http://www.uptodate.com/home/index.html>.
3. UpToDate, Inc. Hypokalemic periodic paralysis. UpToDate [database online]. Waltham, MA. Last updated March 08, 2019 Available at: <http://www.uptodate.com/home/index.html>.
4. Statland JM, Fontaine B, Hanna MG, et al. Review of the Diagnosis and Treatment of Periodic Paralysis. Muscle Nerve. 2018;57(4):522-530.

Policy Implementation/Update:

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| Date Created | September 2015 |
| Date Effective | September 2015 |
| Last Updated | December 2019 |
| Last Reviewed | 12/2019 |

| Action and Summary of Changes | Date |
|---|---------|
| Prior authorization criteria transitioned to policy format. Updated initial and renewal durations as response should be seen within two months of therapy. Addition of specialist requirements. Addition of renewal criteria. | 12/2019 |
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