

## SPECIALTY GUIDELINE MANAGEMENT

### KEVEYIS (dichlorphenamide)

#### POLICY

##### I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

##### FDA-Approved Indication<sup>1</sup>

Treatment of primary hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, and related variants.

All other indications are considered experimental/investigational and are not a covered benefit.

##### II. CRITERIA FOR INITIAL APPROVAL

###### **A. Primary Hypokalemic Periodic Paralysis<sup>1-8</sup>**

Authorization of 60 days may be granted to members who are initiating Keveyis therapy when the following criteria is met:

1. The diagnosis was supported by at least one of the following:
  - a. Genetic test results or
  - b. Patient has a family history of primary hypokalemic periodic paralysis, or
  - c. Patient's attacks are associated with hypokalemia AND both Andersen-Tawil syndrome and thyrotoxic periodic paralysis have been ruled out.
2. Trial with suboptimal response to treatment with acetazolamide

###### **B. Primary Hyperkalemic Periodic Paralysis<sup>1-8</sup>**

Authorization of 60 days may be granted to members who are initiating Keveyis therapy when the following criteria is met:

1. The diagnosis was supported by at least one of the following:
  - a. Genetic test results, or
  - b. Patient has a family history of primary hyperkalemic periodic paralysis, or
  - c. Patient's attacks are associated with hyperkalemia AND Andersen-Tawil syndrome has been ruled out.
2. Trial with suboptimal response to treatment with acetazolamide

##### III. CONTINUATION OF THERAPY

Authorization of 12 months may be granted to members who have demonstrated a response to Keveyis therapy as demonstrated by an improvement in their condition (e.g. decrease in the number or severity of attacks).

#### IV. REFERENCE

1. Keveyis [package insert]. Feasterville-Treose, PA: Strongbridge Biopharma; January 2017.
2. Micromedex Solutions [electronic version]. Truven Health Analytics, Greenwood Village, Colorado, USA. Available at: <http://www.micromedexsolutions.com/>. Accessed February 9, 2018.
3. Lexicomp Online®, Lexi-Drugs®, Hudson, Ohio: Wolters Kluwer Clinical Drug Information, Inc.; <http://online.lexi.com> Accessed February 9, 2018.
4. Clinical Pharmacology [database online]. Atlanta, GA: Elsevier, Inc.; 2018. <https://www.clinicalkey.com/pharmacology/> Accessed February 9, 2018
5. Lexicomp Online®, AHFS® Drug Information, Hudson, Ohio: Wolters Kluwer Clinical Drug Information, Inc.; <http://online.lexi.com> Accessed February 9, 2018.
6. Levitt JO. Practical aspects in the management of hypokalemic periodic paralysis. *Journal of Translational Medicine* 2008; 6:18.
7. Charles G, Zheng C, Lehmann-Horn F, et al. Characterization of hyperkalemic periodic paralysis: a survey of genetically diagnosed individuals. *JNeurol* 2013; 260:2606-2613.
8. Statland, Jeffrey & Fontaine, Bertrand & G. Hanna, Michael & Johnson, Nicholas & T. Kissel, John & A. Sansone, Valeria & Shieh, Perry & N. Tawil, Rabi & Trivedi, Jaya & Cannon, Stephen & C. Griggs, Robert. (2017). A Review of the Diagnosis and Treatment of Periodic Paralysis. *Muscle & Nerve* 10.1002/mus.26009.