Reference number(s)
2491-A

Specialty Guideline Management dichlorphenamide-Keveyis-Ormalvi

Products Referenced by this Document

Drugs that are listed in the following table include both brand and generic and all dosage forms and strengths unless otherwise stated. Over-the-counter (OTC) products are not included unless otherwise stated.

Brand Name	Generic Name
Keveyis	dichlorphenamide
Ormalvi	dichlorphenamide

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 Indications[1-3]

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

All other indications are considered experimental/investigational and not medically necessary.

Coverage Criteria

Primary Hypokalemic Periodic Paralysis[1-6]

Authorization of 60 days may be granted for treatment of primary hypokalemic periodic paralysis when all of the following criteria are met:

- The diagnosis was supported by at least one of the following:

    - Genetic test results, or
    - Member has a family history of primary hypokalemic periodic paralysis, or
    - Member's attacks are associated with hypokalemia AND both Andersen-Tawil syndrome and thyrotoxic periodic paralysis have been ruled out.
- Member had a trial and suboptimal response to treatment with acetazolamide.

Primary Hyperkalemic Periodic Paralysis[1-6]

Authorization of 60 days may be granted for treatment of primary hyperkalemic periodic paralysis when all of the following criteria are met:

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

Continuation of Therapy

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

References

1. Keveyis [package insert]. Chicago, IL: Xeris Pharmaceuticals, Inc.; August 2024.
2. Dichlorphenamide [package insert]. Basking Ridge, NJ: Torrent Pharma Inc.; January 2023.
3. Ormalvi [package insert]. Cambridge, UK: Cycle Pharmaceuticals LTD; February 2024.
4. Levitt JO. Practical aspects in the management of hypokalemic periodic paralysis. J Transl Med. 2008;6:18.
5. Charles G, Zheng C, Lehmann-Horn F, Jurkat-Rott K, Levitt J. Characterization of hyperkalemic periodic paralysis: a survey of genetically diagnosed individuals. J Neurol. 2013;260(10):2606-2613.
6. Statland JM, Fontaine B, Hanna MG, et al. Review of the diagnosis and treatment of periodic paralysis. Muscle Nerve. 2018;57(4):522-530.