Clinical Policy: Dichlorphenamide (Keveyis)

Reference Number: PA.CP.PMN.261
Effective Date: 01/2021
Last Review Date: 01/2024 Revision Log

Description
Dichlorphenamide (Keveyis®) is an oral carbonic anhydrase inhibitor.

FDA Approved Indication(s)
Keveyis is indicated for the treatment of primary hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, and related variants.

Policy/Criteria
Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.

It is the policy of PA Health & Wellness® that Keveyis is **medically necessary** when the following criteria are met:

I. Initial Approval Criteria

A. Hyperkalemic/Hypokalemic Periodic Paralysis and Variants (must meet all)**:**

1. Diagnosis of primary hyperkalemic or hypokalemic periodic paralysis, or related variants (i.e., Andersen's syndrome, paramyotonia congenita);
2. Age ≥ 18 years;
3. Failure of acetazolamide at up to maximally indicated dose, unless contraindicated or clinically significant adverse effects are experienced;
4. If request is for brand Keveyis, member must use generic dichlorphenamide, unless contraindicated or clinically significant adverse effects are experienced;
5. Dose does not exceed 200 mg (4 tablets) per day.

Approval duration: 3 months

B. Other diagnoses/indications

1. Refer to the off-label use policy if diagnosis is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized): PA.CP.PMN.53

II. Continued Therapy

A. Hyperkalemic/Hypokalemic Periodic Paralysis and Variants (must meet all)**:**

1. Currently receiving medication via PA Health & Wellness benefit and documentation supports positive response to therapy or the Continuity of Care policy (PA.LTSS.PHAR.01) applies;
2. Member is responding positively to therapy as evidenced by reduced frequency of paralysis;
3. If request is for brand Keveyis, member must use generic dichlorphenamide, unless contraindicated or clinically significant adverse effects are experienced;
4. If request is for a dose increase, new dose does not exceed 200 mg (4 tablets) per day.

Approval duration: 12 months

B. Other diagnoses/indications (must meet 1 or 2)**:**
1. Currently receiving medication via PA Health & Wellness benefit and documentation supports positive response to therapy or the Continuity of Care policy (PA.LTSS.PHAR.01) applies.
   Approval duration: Duration of request or 6 months (whichever is less); or
2. Refer to the off-label use policy if diagnosis is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized): PA.CP.PMN.53

III. Diagnoses/Indications for which coverage is NOT authorized:

A. Non-FDA approved indications, which are not addressed in this policy, unless there is sufficient documentation of efficacy and safety according to the off label use policies – PA.CP.PMN.53

IV. Appendices/General Information

Appendix A: Abbreviation/Acronym Key
FDA: Food and Drug Administration

Appendix B: Therapeutic Alternatives
This table provides a listing of preferred alternative therapy recommended in the approval criteria. The drugs listed here may not be a formulary agent for all relevant lines of business and may require prior authorization.

Drug Name	Dosing Regimen	Dose Limit/ Maximum Dose
acetazolamide (Diamox®)	250 to 1,000 mg/day PO in divided doses	1,000 mg/day

Therapeutic alternatives are listed as Brand name® (generic) when the drug is available by brand name only and generic (Brand name®) when the drug is available by both brand and generic.

Appendix C: Contraindications/Boxed Warnings
- Contraindication(s): hepatic insufficiency, severe pulmonary obstruction, hypersensitivity to dichlorphenamide or other sulfonamides, concomitant use of Keveyis and high dose aspirin
- Boxed warning(s): none reported

Appendix D: General Information
- Variants of periodic paralysis include paramyotonia congenita and Andersen syndrome.
- Per the Keveyis Prescribing Information: 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 the patient's response to Keveyis after 2 months of treatment to decide whether Keveyis should be continued.

V. Dosage and Administration

Indication	Dosing Regimen	Maximum Dose
Primary hyperkalemic periodic paralysis, primary	Initial dose of 50 mg PO QD or BID; titrate based on individual response at	200 mg/day

Indication	Dosing Regimen	Maximum Dose
hypokalemic periodic paralysis, and related variants	weekly intervals up to a maximum recommended daily dose of 200 mg	

VI. Product Availability

Tablet: 50 mg

VII. References

1. Keveyis Prescribing Information. Hawthorne, NY: Taro Pharmaceuticals U.S.A, Inc.; May 2023. Available at: https://keveyis.com/. Accessed on October 13, 2023.
2. Micromedex® Healthcare Series [Internet database]. Greenwood Village, Colo: Thomson Healthcare. Updated periodically. Accessed November 14, 2023.
3. Tawil R, McDermott MP, Brown R, et al. Randomized trials of dichlorphenamide in the periodic paralyses. Ann Neurol 2000;47:46-53.
4. Venance SL, Cannon SC, Fialho D, et al. The primary periodic paralyses: diagnosis, pathogenesis and treatment. Brain 2006; 129:8.
5. Statland JM, Fontaine B, Hanna MG, et al. Review of the diagnosis and treatment of periodic paralysis. Muscle Nerve 2018;54(4):522-530.

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Reviews, Revisions, and Approvals	Date
Policy created	01/2021
1Q 2022 annual review: references reviewed and updated.	01/2022
1Q 2023 annual review: no significant changes; references reviewed and updated.	01/2023
1Q 2024 annual review: added requirement for use of generic for brand Keveyis requests; references reviewed and updated.	01/2024