Clinical Policy Title:	dichlorphenamide
Policy Number:	RxA.191
Drug(s) Applied:	Keveyis®
Original Policy Date:	02/07/2020
Last Review Date:	06/10/2021
Line of Business Policy Applies to:	All lines of business

Background

Dichlorphenamide is an oral carbonic anhydrase inhibitor. It is indicated for primary hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, and related variants.

Dosing Information

Drug Name	Indication	Dosing Regimen	Maximum Dose
dichlorphenamide (Keveyis®)	Primary hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, and related variants	Initial dose of 50 mg PO once or twice daily; titrate based on individual response at weekly intervals up to a maximum daily dose of 200 mg.	200 mg/day

Dosage Forms

- Tablet: 50 mg

Clinical Policy

Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria. The provision of provider samples does not guarantee coverage under the terms of the pharmacy benefit administered by RxAdvance. All criteria for initial approval must be met in order to obtain coverage.

I. Initial Approval Criteria

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

1. Member has a diagnosis of primary hyperkalemic or hypokalemic periodic paralysis, or related variants (i.e., Andersen-Tawil syndrome, paramyotonia congenita);
2. Prescribed by or in consultation to a neurologist or provider who specialized in the treatment of primary periodic paralysis (e.g., muscle disease specialist or physiatrist);
3. Member is 18 years of age or older;
4. Failure of acetazolamide at up to maximally indicated dose, unless contraindicated or clinically significant adverse effects are experienced;
5. Dose does not exceed 200 mg (4 tablets) per day.

Approval Duration

Commercial: 2 months

Medicaid: 2 months

II. Continued Therapy Approval

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

1. Member is currently receiving medication that has been authorized by RxAdvance or member has previously met initial approval criteria listed in this policy;
2. Member is responding positively to therapy as evidenced by reduced frequency of attacks;
3. If request is for a dose increase, new dose does not exceed 200 mg (4 tablets) per day.

Approval Duration
Commercial: 12 months
Medicaid: 12 months

III. Appendices

APPENDIX A: Abbreviation/Acronym Key
FDA: Food and Drug Administration
PO: By mouth

APPENDIX B: Therapeutic Alternatives
Below are suggested therapeutic alternatives based on clinical guidance. Please check drug formulary for preferred agents and utilization management requirements.

Drug Name	Dosing Regimen	Dose Limit/ Maximum Dose
acetazolamide	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):
  - Concomitant use of dichlorphenamide and high dose aspirin
  - Hepatic insufficiency
  - Severe pulmonary obstruction
  - Hypersensitivity to dichlorphenamide or other sulfonamides

- Boxed Warning(s):
  - None reported

APPENDIX D: General Information

- Variants of periodic paralysis include paramyotonia congenita and Andersen-Tawil syndrome.
- Primary hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, and related variants are a heterogeneous group of conditions, for which the response to dichlorphenamide may vary. Therefore, prescribers should evaluate the patient's response to dichlorphenamide after 2 months of treatment to decide whether dichlorphenamide should be continued.

References

1. Keveyis Prescribing Information. Trevose, PA: Strongbridge US, Inc.; November 2019. Available at https://www.keveyis.com/wp-content/uploads/keveyis-prescribing-information.pdf . Accessed on March 4, 2021.
2. Tawil R, McDermott MP, Brown R, et al. Randomized trials of dichlorphenamide in the periodic paralyses. Ann Neurol 2000;47:46-53.
3. Venance SL, Cannon SC, Fialho D, et al. The primary periodic paralyses: diagnosis, pathogenesis and treatment. Brain 2006; 129:8.
4. Sansone VA, Burge J, McDermott MP, et al. Randomized, placebo-controlled trials of dichlorphenamide in periodic paralysis. Neurology. 2016; 86: 1408-1416.
5. Statland JM, Fontaine B, Hanna MG, et al. Review of the Diagnosis and Treatment of Periodic Paralysis. Muscle Nerve. 2018; 57(4): 522-530.
6. Micromedex® Healthcare Series [Internet database]. Greenwood Village, Colo: Thomson Healthcare. Updated periodically. Accessed on March 4, 2021.

Review/Revision History	Review/Revision Date	P&T Approval Date
Policy established.	01/2020	02/07/2020
Policy was reviewed 1. Policy title was updated. 2. Lines of Business Policy applies to was updated to all lines of business. 3. Initial and Continued therapy approval duration was updated to include Commercial & Medicaid approval duration. 4. Continued therapy criteria II.A.1 was rephrased to "Currently receiving medication that has been authorized by RxAdvance.." 5. References were updated.	07/07/2020	09/14/2020
Policy was reviewed 1. Policy title was updated. 2. Initial criteria for approval and duration updated. 3. Continued therapy criteria II.A.1 was rephrased to "Member is currently receiving medication that has been authorized by RxAdvance.." 4. Contraindications were updated. 5. References were reviewed and updated.	03/04/2021	6/10/2021