Commercial/Healthcare Exchange PA Criteria

Effective: April 27,2020

Prior Authorization: Keveyis (dichlorphenamide)

Products Affected: Keveyis (dichlorphenamide) oral tablet

Medication Description:
Keveyis, a carbonic anhydrase inhibitor that is FDA indicated for the treatment of primary hyperkalemic periodic paralysis (HyperPP), primary hypokalemic periodic paralysis (HypoPP), and related variants. Periodic paralysis is a rare neuromuscular disorder, related to a defect in muscle ion channels, characterized by episodes of painless muscle weakness, which may be precipitated by heavy exercise, fasting, or high-carbohydrate meals. Periodic paralysis (PP) is classified as hypokalemic when episodes occur in association with low potassium blood levels or as hyperkalemic when episodes can be induced by elevated potassium. Most cases of periodic paralysis are hereditary, usually with an autosomal dominant inheritance pattern. Acquired cases of hypokalemic PP have been described in association with hyperthyroidism. Hypokalemic periodic paralysis (PP) is the most common of the periodic paralyses, but is still quite rare, with an estimated prevalence of 1 in 100,000. Hypokalemic PP may be familial with autosomal dominant inheritance or may be acquired in patients with thyrotoxicosis. Clinical penetrance is often incomplete, especially in women. The disorder is three to four times more commonly clinically expressed in men. Approximately one-third of cases represent new mutations.

The recommended initial dose of Keveyis is 50 mg twice daily (BID). The maximum recommended total daily dose is 200 mg. Primary HyperPP, primary HypoPP, 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 to decide whether it should be continued.

Covered Uses:

1. Hyperkalemic Periodic Paralysis (HyperPP) and Related Variants
2. Hypokalemic Periodic Paralysis (HypoPP) and Related Variants

Exclusion Criteria:
Keveyis has not been shown to be effective, or there are limited or preliminary data or potential safety concerns that are not supportive of general approval in the following circumstances.

1. Patient with history of hypersensitivity to diclorphenamide or other sulfonamides
2. Patient on high dose aspirin
3. Patient with severe pulmonary disease
4. Patient with hepatic insufficiency

Required Medical Information:
1. Diagnosis
2. Potassium concentration during paralytic attack (if available)

3. Pertinent family history related to condition (if available)
4. Pertinent confirmation of skeletal muscle calcium or sodium channel mutation (if available)

Age Restrictions: None

Prescriber Restrictions: Prescribed by or in consultation with a neurologist or a physician who specializes in the care of patients with primary periodic paralysis (e.g., muscle disease specialist, physiatrist).

Coverage Duration:
1. Initial therapy – 2 months
2. Continuing therapy – 12 months

Other Criteria:

1. Hyperkalemic Periodic Paralysis (HyperPP) and Related Variants

A) Patient has a confirmed diagnosis of primary hyperkalemic periodic paralysis by meeting at least ONE of the following criteria (a, b, c, or d):
   a) Patient has had an increase from baseline in serum potassium concentration of greater than or equal to 1.5 mEq/L during a paralytic attack; OR
   b) Patient has had a serum potassium concentration during a paralytic attack of greater than 5.0 mEq/L; OR
   c) Patient has a family history of the condition; OR
   d) Patient has a genetically confirmed skeletal muscle sodium channel mutation; AND

B) The prescribing physician has excluded other reasons for acquired hyperkalemia (e.g., drug abuse, renal and adrenal dysfunction)

Continuation of treatment

A) Patient has decrease in the frequency or severity of paralytic attacks with treatment as determined by the prescribing physician

2. Hypokalemic Periodic Paralysis (HypoPP) and Related Variants

Initiation of treatment

A) Patient has a confirmed diagnosis of primary hypokalemic periodic paralysis by meeting at least ONE of the following (a, b, or c):
   a) Patient has had a serum potassium concentration of less than 3.5 mEq/L during a paralytic attack; OR
   b) Patient has a family history of the condition; OR
   c) Patient has a genetically confirmed skeletal muscle calcium or sodium channel mutation; AND

B) Patient has had improvements in paralysis attack symptoms with potassium intake; AND

Continuation of treatment

B) Patient has decrease in the frequency or severity of paralytic attacks with treatment as determined by the prescribing physician

References:

1. Keveyis™ tablets [prescribing information]. Hawthorne, NY: Taro Pharmaceuticals; August 2015.
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3. Genetics Home Reference. Hypokalemic periodic paralysis. Reviewed April 2007. Available at: http://ghr.nlm.nih.gov/condition/hypokalemic-periodic-paralysis. Accessed on September 15, 2015.
4. Vicart S, Sternberg D, Arzel-Hezode M, et al. Hypokalemic periodic paralysis. Initial posting April 30, 2002. Updated July 31, 2014. GeneReviews® - NCBI Bookshelf. Available at: http://www.ncbi.nlm.nih.gov/books/NBK1338/?report=printable. Accessed on August 12, 2015.
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7. Vicart S, Sternberg D, Arzel-Hezode M, et al. Hypokalemic periodic paralysis. Initial posting April 30, 2002. Updated July 31, 2014. GeneReviews® - NCBI Bookshelf. Available at: http://www.ncbi.nlm.nih.gov/books/NBK1338/?report=printable. Accessed on August 12, 2015.
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Policy Revision history

Rev #	Type of Change	Summary of Change	Sections Affected	Date
1	New Policy	New Policy	All	1/13/2016
2	Update	CCI adoption of EH policy Removal from CCI Pa to Indication Policy	Reviewed covered uses and exclusion criteria to FDA label	4/27/2020