Drug Name: Keveyis (dichlorphenamide)
Effective Date: 8/1/2019
Revised:
Scope: Medicaid

Drug Name:	Keveyis (dichlorphenamide)
Required Medical Information:	***Initial Coverage Criteria:*** • Must be prescribed by a neurologist; AND • Must have a diagnosis of primary hyperkalemic periodic paralysis, primary hypokalemic periodic paralysis, or related variants; AND • Must not have any of the following: o Concomitant use with high dose aspirin o Severe pulmonary disease, limiting compensation to metabolic acidosis that may be caused by dichlorphenamide (Keveyis) o Hepatic encephalopathy; AND **Primary hypokalemic periodic paralysis**: • Must have documentation confirming diagnosis, defined as one of the following scenarios: o Two or more attacks of muscle weakness with documented serum K <3.5mEq/L, OR o One attack of muscle weakness in the member with documented serum potassium < 3.5mEq/L and one attack of weakness in a relative with a history of the condition, OR o Three of the following clinical/laboratory features: ▪ Onset of symptoms in the first or second decade of life ▪ Duration of attack (muscle weakness involving one or more limbs) longer than two hours ▪ The presence of triggers (previous carbohydrate rich meal, symptom onset during rest after exercise or during stressful situations) for attacks ▪ Improvement in symptoms with potassium intake ▪ A family history of the condition or genetically confirmed skeletal calcium or sodium channel mutation ▪ Positive long exercise test (LET: recording evoked compound motor action potentials (CMAP) over 30-45 minutes, every one to two minutes and then every five minutes, after long

effort (2-5 minutes, with brief 3 to 4 second rest periods every 15-45 seconds).

- Must have chart documentation excluding other causes of hypokalemia (renal, adrenal, thyroid dysfunction; renal tubular acidosis; diuretic and laxative abuse); AND
- Must have an adequate trial and failure or an inadequate response of acetazolamide unless has one of the following mutations in SCNA4:
  - c.2014C>G
  - c.2014C>A; AND
- Must be using a potassium supplement: AND
- Must have been counseled on appropriate dietary restrictions (high potassium intake, low sodium intake, and low carbohydrate intake)

Hyperkalemic periodic paralysis:

- Must have documentation confirming diagnosis::
  - For diagnosis confirmed by genetic testing, must have both of the following:
    - A family history of the condition or genetically confirmed skeletal sodium channel mutation associated with hyperkalemic periodic paralysis, AND
    - A history of at least two attacks of flaccid limb weakness (which may also include weakness of the muscles of the eyes, throat, and trunk) or 1 attack with a family history of attacks of hyperkalemic periodic paralysis;

      OR
  - For diagnosis made based on clinical presentation, must have all of the following:
    - A history of at least two attacks of flaccid limb weakness (which may also include weakness of the muscles of the eyes, throat, and trunk) or 1 attack with a family history of attacks of hyperkalemic periodic paralysis
    - Serum potassium >5mEq/L or an increase of serum potassium concentration of at least 1.5 mEq/L during an attack of weakness and/or onset/worsening of an attack as a result of oral potassium intake
    - Presence of myotonia OR any 3 of the following clinical features:
      - Typical attack duration less than 2

hours
- Onset before 30 years.
- Positive long exercise test (>40% decrement in CMAP)
- Typical external triggers (rest after exercise, potassium load, fasting)

- Must have chart documentation of normal serum potassium concentration and muscle strength between attacks; AND
- Must have chart documentation of ECG recording for the exclusion of a long QT and ventricular arrhythmias; AND
- Must not have either of the following:
  - Secondary hyperkalemic periodic paralysis due to ingestion of potassium or of a potassium sparing diuretic
  - Paramyotonia (i.e. muscle stiffness that is worsening after exercise or cold-induced); AND
- Must have documentation of exclusion of other hereditary forms of hyperkalemia (i.e., Andersen-Tawil syndrome) and acquired forms of hyperkalemia (drug abuse, renal and adrenal dysfunction):AND
- Must have an adequate trial of acetazolamide with an inadequate response; AND
- Must have been counseled on appropriate dietary recommendations(such as frequent meals high in carbohydrate)

Renewal Criteria:

- Patient is tolerating treatment; AND
- Clinical documentation from the prescriber that the member's condition has improved based upon the prescriber's assessment while on therapy

Approval durations:
Initial: 2 months
Renewal: 12 months

Quantity Limit of 200mg per day (120 tablets per 25 days)