Oscar Clinical Guideline: Relyvrio (Sodium Phenylbutyrate/Taurursodiol) (PG129, Ver. 1)

Relyvrio (Sodium Phenylbutyrate/Taurursodiol)

Disclaimer

Clinical guidelines are developed and adopted to establish evidence-based clinical criteria for utilization management decisions. Clinical guidelines are applicable according to policy and plan type. The Plan may delegate utilization management decisions of certain services to third parties who may develop and adopt their own clinical criteria.

Coverage of services is subject to the terms, conditions, and limitations of a member's policy, as well as applicable state and federal law. Clinical guidelines are also subject to in-force criteria such as the Centers for Medicare & Medicaid Services (CMS) national coverage determination (NCD) or local coverage determination (LCD) for Medicare Advantage plans. Please refer to the member's policy documents (e.g., Certificate/Evidence of Coverage, Schedule of Benefits, Plan Formulary) or contact the Plan to confirm coverage.

Summary

Amyotrophic lateral sclerosis (ALS) is a disease that affects voluntary muscle control and movement due to nerve damage to neurons (nerve cells) in the brain and spinal cord. ALS is sometimes also referred to as motor neuron disease, classic motor neuron disease, Lou Gehrig disease, or Charcot disease. ALS gets worse over time, leading to death within three to five years for most people. Early in the disease, symptoms may include cramping, muscle twitches/spasms, weakness in arms or legs, difficulty chewing/swallowing, or slurred speech. As the disease progresses, signs and symptoms worsen - leading to partial or total paralysis and eventually, death.

ALS is primarily a clinical diagnosis identified with history and physical examination with use of classification/staging criteria. Other tests (e.g., electromyogram, nerve conduction studies, imaging studies such as an MRI or a CT scan) may help confirm the diagnosis by ruling out conditions that can mimic amyotrophic lateral sclerosis. Around roughly 10% of people with ALS are thought to have an inherited (referred to as familial ALS) form of the disease, with the remainder affected by a sporadic (non-inherited) form.

Non-drug treatments may include physical therapy, occupational therapy, speech therapy, breathing support (devices), and a feeding tube. Medicines may be used to treat muscle spasms and weakness, drooling, sleep problems, pain, anxiety, depression, or pseudobulbar affect. There is currently no cure for ALS, but treatments are available that may help with symptoms or to slow the progress of the disease to help people live longer such as riluzole (Rilutek), edaravone (Radicava), and sodium phenylbutyrate and taurursodiol (Relyvrio).

Definitions

"Amyotrophic lateral sclerosis (ALS)", is a disease that affects voluntary muscle control and movement due to nerve damage to neurons (nerve cells) in the brain and spinal cord. ALS is sometimes also referred to as motor neuron disease, classic motor neuron disease, Lou Gehrig disease, or Charcot disease.

"Atrophy", is the wasting, or progressive loss of muscle mass due to reduction in the size or number of muscle cells.

"Central Nervous System (CNS)", is the brain and spinal cord.

"Dementia", is a condition in which memory and thinking are affected.

"Frontotemporal dementia", is a type of dementia caused by damage to nerve cells in certain parts of the brain. ALS worsens over time and may cause frontotemporal dementia.

"Neurologist", is a physician who specializes in the nervous system and its disorders.

"Neurotransmitter" is a molecule that sends signals from neurons to different parts of the body (e.g., muscles).

Medical Necessity Criteria for Initial Authorization

The Plan considers Relyvrio (Sodium Phenylbutyrate/Taurursodiol) medically necessary when **ALL** of the following criteria are met:

1. Prescribed by or in consultation with a neurologist or physician specializing in the treatment of amyotrophic lateral sclerosis (ALS); ***AND***
2. The member is 18 years of age or older; ***AND***

3. The member has a diagnosis of amyotrophic lateral sclerosis (ALS) AND documentation has been provided showing BOTH of the following:
    a. Comprehensive assessment of clinical signs and symptoms, with electrodiagnostic testing and neuroimaging to exclude other conditions in differential diagnosis; ***and***
    b. The diagnosis of ALS has been confirmed by a second opinion from another neurologist or physician specializing in the treatment of amyotrophic lateral sclerosis (ALS); ***AND***
4. Is being prescribed within the manufacturer's published dosing guidelines or falls within dosing guidelines found in a compendia of current literature (i.e., 1 packet twice daily); ***AND***
5. Clinical chart documentation is provided for review to substantiate the above listed requirements.

If the above prior authorization criteria are met, Relyvrio (Sodium Phenylbutyrate/Taurursodiol) will be approved for 12 months.

Medical Necessity Criteria for Reauthorization

Reauthorization for 12 months will be granted if BOTH of the following are met:

1. the member still meets the applicable initial criteria***; AND***
2. recent chart documentation (within the last 12 months) shows the member has experienced a positive clinical benefit since starting the requested medication as evidenced by a positive effect on the rate of reduction of fine motor, gross motor, bulbar, and respiratory functions decline (i.e., slowing of the decline of functional abilities such as speech, salivation, swallowing, handwriting, cutting food, dressing/hygiene, turning in bed, walking, climbing stairs, dyspnea, orthopnea, and respiratory insufficiency).

Experimental or Investigational / Not Medically Necessary

Relyvrio (Sodium Phenylbutyrate/Taurursodiol) for any other indication is considered not medically necessary by the Plan, as it is deemed to be experimental, investigational, or unproven.

Table 1: ICD-10-CM (diagnosis) Codes for Migraine and Cluster Headaches

Codes	Description
G12.21	Amyotrophic lateral sclerosis

References

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Clinical Guideline Revision / History Information

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