MEDICAL MUTUAL®

Drug Policy

Policy:	Relyvrio (sodium phenylbutyrate/taurursodiol) oral packets	Annual Review Date: 05/18/2023 Last Revised Date: 05/18/2023

OVERVIEW

Relyvrio, a combination product of sodium phenylbutyrate and taurursodiol, is indicated for the treatment of **amyotrophic lateral sclerosis** (ALS) in adults.[1]

POLICY STATEMENT

This policy involves the use of Relyvrio. Prior authorization is recommended for pharmacy benefit coverage of Relyvrio. Approval is recommended for those who meet the conditions of coverage in the **Criteria and Initial/Extended Approval** for the diagnosis provided. **Conditions Not Recommended for Approval** are listed following the recommended authorization criteria. Requests for uses not listed in this policy will be reviewed for evidence of efficacy and for medical necessity on a case-by-case basis.

Because of the specialized skills required for evaluation and diagnosis of patients treated with Relyvrio as well as the monitoring required for adverse events and long-term efficacy, initial approval requires Relyvrio be prescribed by or in consultation with a physician who specializes in the condition being treated. All approvals for initial therapy are provided for the initial approval duration noted below; if reauthorization is allowed, a response to therapy is required for continuation of therapy unless otherwise noted below.

RECOMMENDED AUTHORIZATION CRITERIA

Coverage of Relyvrio is recommended in those who meet the following criteria:

1. **Amyotrophic Lateral Sclerosis (ALS).** Approve for duration noted below if the patient meets ONE of the following (A or B):
   - **A)** Initial Therapy. Approve if the patient meets ALL the following (i, ii, iii, iv, v, vi, vii, and viii):
     - **i.** Patient is ≥ 18 years of age; AND
     - **ii.** According to the prescriber, the patient has a "definite" diagnosis of amyotrophic lateral sclerosis (ALS) based on the application of the revised El Escorial/revised Airlie House criteria or Awaji-Shima criteria; AND
     - **iii.** Patient does not have a tracheostomy or permanent assisted ventilation (PAV); AND
     - **iv.** Patient has a percent-predicted slow vital capacity (SVC) > 60% based on gender, height, and age; AND
     - **v.** Onset of ALS symptoms began within the preceding 18 months; AND
     - **vi.** Patient meets one of the following (i, ii, or iii):
       - **i.** Patient has previously received a riluzole product; OR
       - **ii.** Patient is currently receiving a riluzole product; OR

iii. Patient will take Relyvrio concomitantly with a riluzole product; AND
Note: Examples of riluzole products include riluzole tablets, Tiglutik (riluzole oral suspension), or Exservan (riluzole oral film).

vii. Patient will not use Relyvrio concomitantly with any other medications containing phenylbutyrate or taurursodiol, including over-the-counter supplements; AND

viii. The medication is prescribed by or in consultation with a neurologist, a neuromuscular disease specialist, or a physician specializing in the treatment of ALS.

B) Patient is Currently Receiving Relyvrio. Approve if the patient meets ALL of the following (i, ii, iii, iv, v, and vi):

i. Patient is ≥ 18 years of age; AND

ii. Patient does not have a tracheostomy; AND

iii. Patient does not require permanent assisted ventilation (> 22 hours per day); AND

iv. Patient will not use Relyvrio concomitantly with any other medications containing phenylbutyrate or taurursodiol, including over-the-counter supplements; AND

v. According to the prescriber, the patient has experienced slowed disease progression from baseline; AND

vi. The medication is prescribed by or in consultation with a neurologist, a neuromuscular disease specialist, or a physician specializing in the treatment of ALS.

Initial Approval/ Extended Approval.
A) *Initial Approval:* 6 months
B) *Extended Approval:* 6 months

CONDITIONS NOT RECOMMENDED FOR APPROVAL

Relyvrio 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 for the following conditions. (Note: This is not an exhaustive list of Conditions Not Recommended for Approval).

1. Coverage is not recommended for circumstances not listed in the Recommended Authorization Criteria. Criteria will be updated as new published data are available.

Documentation Requirements:

The Company reserves the right to request additional documentation as part of its coverage determination process. The Company may deny reimbursement when it has determined that the drug provided or services performed were not medically necessary, investigational or experimental, not within the scope of benefits afforded to the member and/or a pattern of billing or other practice has been found to be either inappropriate or excessive. Additional documentation supporting medical necessity for the services provided must be made available upon request to the Company. Documentation requested may include patient records, test results and/or credentials of the provider ordering or performing a service. The Company also reserves the right to modify, revise, change, apply and interpret this policy at its sole discretion, and the exercise of this discretion shall be final and binding.

REFERENCES

1. Relyvrio™ oral suspension [prescribing information]. Cambridge, MA: Amylyx; September 2022.
2. Paganoni S, Macklin EA, Hendrix S, et al. Trial of sodium phenylbutyrate-taurursodiol for amyotrophic lateral sclerosis. *N Engl J Med*. 2020 Sep 3;383(10):919-930.
3. Miller RG, Jackson CE, Kasarskis EJ, et al. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: multidisciplinary care, symptom management, and cognitive/behavioral impairment (an evidence-based review). *Neurology*. 2009 (reaffirmed 2020);73(15):1227-1233.
4. Miller RG, Jackson CE, Kasarskis EJ, et al. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review). *Neurology*. 2009;73:1218-1226.
5. Andersen PM, Abrahams S, Borasio GD, et al. EFNS guidelines on the clinical management of amyotrophic lateral sclerosis (MALS) – revised report of an EFNS task force. *Eur J Neurol*. 2012;19(3):360-375.