| Reference number(s) |
| --- |
| 5632-A |

# Specialty Guideline Management Relyvrio

## Products Referenced by this Document

Drugs that are listed in the following table include both brand and generic and all dosage forms and strengths unless otherwise stated. Over-the-counter (OTC) products are not included unless otherwise stated.

| Brand Name | Generic Name |
| --- | --- |
| Relyvrio | sodium phenylbutyrate and taurursodiol |

## Indications

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

### FDA-Approved Indications[1]

Relyvrio is indicated for the treatment of amyotrophic lateral sclerosis (ALS) in adults.

All other indications are considered experimental/investigational and not medically necessary.

## Documentation

Submission of the following information is necessary to initiate the prior authorization review:

Chart notes, medical record documentation, and/or laboratory results supporting use as applicable in the coverage criteria and continuation of therapy sections.

- Initial Requests:
  - Diagnosis of probable or definite amyotrophic lateral sclerosis (ALS).
  - Forced Vital Capacity (FVC) or Slow Vital Capacity (SVC) results.

- Continuation Requests:
  - Documentation of clinical benefit from therapy with the requested medication.

# Prescriber Specialties

This medication must be prescribed by or in consultation with a neurologist, neuromuscular specialist, or physician specializing in the treatment of amyotrophic lateral sclerosis (ALS).

# Coverage Criteria

## Amyotrophic Lateral Sclerosis (ALS)[1-5]

Authorization of 12 months may be granted for treatment of amyotrophic lateral sclerosis (ALS) when all of the following criteria are met:

- Member has a diagnosis of probable or definite ALS (e.g., medical history and/or diagnostic testing including, nerve conduction studies, imaging and laboratory values to support the diagnosis).
- Member is 18 years of age or older.
- Member has a forced vital capacity (FVC) > 60% or slow vital capacity (SVC) > 60% of predicted value for gender, height, and age.
- Member does not have a tracheostomy.

# Continuation of Therapy

Authorization of 12 months may be granted for members requesting continuation of therapy when all of the following criteria are met:

- Member has a diagnosis of probable or definite amyotrophic lateral sclerosis (ALS).
- Member has a documented clinical benefit from therapy with the requested medication.
- Invasive ventilation or tracheostomy is not required.

# References

1. Relyvrio [package insert]. Cambridge, MA: Amylyx Pharmaceuticals, Inc.; September 2022.
2. EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis; Andersen PM, 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):360-75.

3. Paganoni S, Macklin EA, Hendrix S, et al. Trial of sodium phenylbutyrate-taurursodiol for amyotrophic lateral sclerosis. N Engl J Med 2020; 383:919-30.
4. Paganoni S, Hendrix S, Dickson SP, et al. Long-term survival of participants in the CENTAUR trial of sodium phenylbutyrate-taurursodiol in amyotrophic lateral sclerosis. Muscle Nerve 2021; 63:31-9.
5. Paganoni S, Hendrix S, Dickson SP, et al. Effect of sodium phenylbutyrate/taurursodiol on tracheostomy/ventilation-free survival and hospitalization in amyotrophic lateral sclerosis: long-term results from the CENTAUR trial. J Neurol Neurosurg Psychiatry 2022.
6. 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; 73(15):1227-1233.
7. Pinto S, de Carvalho M. Correlation between Forced Vital Capacity and Slow Vital Capacity for the assessment of respiratory involvement in Amyotrophic Lateral Sclerosis: a prospective study. Amyotrophic lateral sclerosis & frontotemporal degeneration. 2017;18(1-2):86-91.