Reference number
5632-A

SPECIALTY GUIDELINE MANAGEMENT

RELYVRIO (sodium phenylbutyrate and taurursodiol)

POLICY

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

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

II. DOCUMENTATION

Submission of the following information is necessary to initiate the prior authorization review:
Chart notes or medical record documentation supporting use as applicable in section IV and V.

A. Initial Requests:
   1. Diagnosis of probable or definite ALS (e.g., medical history and diagnostic testing including, nerve conduction studies, imaging and laboratory values to support the diagnosis)
   2. Forced Vital Capacity (FVC) > 60% or Slow Vital Capacity (SVC) >60% of predicted value for gender, height, and age

B. Continuation Requests:
   1. Documentation of clinical benefit from Relyvrio therapy

III. 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).

IV. CRITERIA FOR INITIAL APPROVAL

Authorization of 12 months may be granted for treatment of ALS when all of the following criteria are met:

A. Diagnosis of probable or definite ALS (e.g., medical history and diagnostic testing including, nerve conduction studies, imaging and laboratory values to support the diagnosis)
B. Member is 18 years of age or older
C. Forced Vital Capacity (FVC) > 60% or Slow Vital Capacity (SVC) >60% of predicted value for gender, height, and age
D. Member does not have a tracheostomy

V. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for members continuing with Relyvrio therapy for the treatment of ALS when the following criteria are met:

A. Diagnosis of probable or definite ALS
B. There is a clinical benefit from Relyvrio therapy
C. Invasive ventilation or tracheostomy is not required

VI. 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.
8. 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.