BlueCross BlueShield
Federal Employee Program
Blue Cross Blue Shield Association
750 9th St NW, Suite 900
Washington, D.C. 20001
1-800-624-5060
Fax 1-877-378-4727

5.75.038

Section:	Prescription Drugs	**Effective Date:**	January 1, 2025
Subsection:	Neuromuscular Agents	**Original Policy Date:**	October 28, 2022
Subject:	Relyvrio	**Page:**	1 of 4

Last Review Date: December 13, 2024

Relyvrio

Description

Relyvrio (sodium phenylbutyrate and taurursodiol) powder for suspension

Background

Relyvrio (sodium phenylbutyrate and taurursodiol) is a powder for suspension indicated for the treatment of patients with amyotrophic lateral sclerosis (ALS) (1). The mechanism by which this happens is not clearly known but the combination of sodium phenylbutyrate and taurursodiol may decrease neuronal cell death through reduction of endoplasmic reticulum stress and mitochondrial dysfunction (2).

Regulatory Status

FDA-approved indication: Relyvrio is indicated for the treatment of amyotrophic lateral sclerosis (ALS) in adults (1).

In patients with disorders interfering with bile acid circulation, absorption of either components of Relyvrio may be decreased. Monitor for new or worsening diarrhea and consider consulting with a specialist (1).

Relyvrio has a high sodium content. In patients sensitive to sodium intake, consider the amount of sodium in each dose of Relyvrio and monitor as appropriate (1).

Studies have shown that riluzole is safe and effective for slowing disease progression to a modest degree in ALS. Riluzole is considered first-line therapy along with nutritional supplements for patients with ALS (3).

The safety and effectiveness of Relyvrio in pediatric patients less than 18 years of age have not been established (1).

Related policies
Exservan, Qalsody, Radicava

Policy

This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.

Relyvrio may be considered **medically necessary** and if the conditions indicated below are met.

Relyvrio may be considered **investigational** for all other indications.

Prior-Approval Requirements

Age 18 years of age and older

Diagnosis

Patient must have the following:

Amyotrophic lateral sclerosis (ALS)

AND ALL of the following:

1. Patient has had an inadequate treatment response to riluzole or will continue to take riluzole
2. Prescriber agrees to monitor patient's sodium intake, if appropriate (e.g., in patients with heart failure, hypertension, or renal impairment)
3. Prescribed by or recommended by a neurologist

Prior – Approval *Renewal* Requirements

Age 18 years of age and older

Diagnosis

Patient must have the following:

Amyotrophic lateral sclerosis (ALS)

AND ALL of the following:

1. Documented stabilization, slowing of disease progression, or improvement of the condition
2. Prescriber agrees to monitor patient's sodium intake, if appropriate (e.g., in patients with heart failure, hypertension, or renal impairment)
3. Prescribed by or recommended by a neurologist

Policy Guidelines

Pre - PA Allowance

None

Prior - Approval Limits

Quantity 168 single-dose packets per 84 days

Duration 12 months

Prior – Approval *Renewal* Limits

Same as above

Rationale

Summary

Relyvrio (sodium phenylbutyrate and taurursodiol) is a powder for suspension indicated for the treatment of patients with amyotrophic lateral sclerosis (ALS). Relyvrio has a high sodium content. In patients sensitive to sodium intake, the amount of sodium in each dose of Relyvrio should be considered and the patient should be monitored as appropriate. The safety and effectiveness of Relyvrio in pediatric patients less than 18 years of age have not been established (1).

Prior authorization is required to ensure the safe, clinically appropriate, and cost-effective use of Relyvrio while maintaining optimal therapeutic outcomes.

References

1. Relyvrio [package insert]. Cambridge MA: Amylyx Pharmaceuticals; September 2022.
2. Sodium phenylbutyrate and taurursodiol. Mechanism of Action. Clinical Pharmacology. Accessed on July 13, 2023.
3. Miller R, Jackson C, et al. Practice Parameter update: The care of the patient with amyotrophic lateral sclerosis: Drug, nutritional, and respiratory therapies (an evidence-based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology. Neurology 2009; 73; 1218-1226

Policy History

Date	Action
October 2022	Addition to PA
December 2022	Annual review
March 2023	Annual review
September 2023	Annual review and reference update
September 2024	Annual review
December 2024	Annual review

Keywords

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on December 13, 2024 and is effective on January 1, 2025.