COMMON GROUND
HEALTHCARE COOPERATIVE

PHARMACY POLICY STATEMENT
Common Ground Healthcare Cooperative (CGHC)

DRUG NAME	**Relyvrio (sodium phenylbutyrate/ taurursodiol)**
BILLING CODE	Must use valid NDC
BENEFIT TYPE	Pharmacy
STATUS	Prior Authorization Required

Relyvrio is an oral, fixed-dose combination of sodium phenylbutyrate and taurursodiol approved by the FDA in 2022, for the treatment of amyotrophic lateral sclerosis (ALS). It is designed to reduce neuronal death by simultaneously mitigating endoplasmic reticulum stress and mitochondrial dysfunction. Approval was based on data from the phase 2 CENTAUR study and its open label extension (CENTAUR-OLE). Relyvrio may be prescribed alone or in combination with other disease modifying therapies, riluzole and/or edaravone, all having different mechanisms of action. ALS is a rare neurodegenerative disease characterized by progressive muscle weakness. Cognitive dysfunction or dementia may also occur. Respiratory failure is the predominant cause of death.

Relyvrio (sodium phenylbutyrate/ taurursodiol) will be considered for coverage when the following criteria are met:

Amyotrophic Lateral Sclerosis (ALS)

For **initial** authorization:

1. Member is at least 18 years of age; AND
2. Medication must be prescribed by or in consultation with a neurologist; AND
3. Member has documentation of ALS diagnosed as definite as defined by the revised El Escorial criteria (clinical evidence of both upper and lower motor neuron signs in at least three body regions); AND
4. ALS symptom onset was within the last 18 months; AND
5. Member's slow vital capacity (SVC) is >60% of predicted value; AND
6. Member does NOT have tracheostomy or permanent assisted ventilation (PAV).
7. **Dosage allowed/Quantity limit:**
   Initially, 1 packet (3 g sodium phenylbutyrate and 1 g taurursodiol powder for oral suspension) daily for the first 3 weeks. After 3 weeks, increase to the maintenance dosage of 1 packet twice daily.
   QL: 56 packets per 28 days

If all the above requirements are met, the medication will be approved for 6 months.

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For **reauthorization**:
1. Chart notes must include documentation of slowed rate of functional decline (may be measured via the ALSFRS-R scale).

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Relyvrio (sodium phenylbutyrate/ taurursodiol) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
11/14/2022	New policy for Relyvrio created.

References:

1. Relyvrio [prescribing information]. Amylyx Pharmaceuticals, Inc., 2022.
2. Paganoni S, Macklin EA, Hendrix S, et al. Trial of Sodium Phenylbutyrate-Taurursodiol for Amyotrophic Lateral Sclerosis. *N Engl J Med*. 2020;383(10):919-930. doi:10.1056/NEJMoa1916945
3. 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(1):31-39. doi:10.1002/mus.27091
4. Paganoni S, Hendrix S, Dickson SP, et al. Effect of sodium phenylbutyrate/taurursodiol on tracheostomy/ventilation-free survival and hospitalisation in amyotrophic lateral sclerosis: long-term results from the CENTAUR trial [published online ahead of print, 2022 May 16]. *J Neurol Neurosurg Psychiatry*. 2022;93(8):871-875. doi:10.1136/jnnp-2022-329024
5. Elia AE, Lalli S, Monsurrò MR, et al. Tauroursodeoxycholic acid in the treatment of patients with amyotrophic lateral sclerosis [published correction appears in Eur J Neurol. 2017 Apr;24(4):659]. *Eur J Neurol*. 2016;23(1):45-52. doi:10.1111/ene.12664
6. EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis:, Andersen PM, Abrahams S, 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. doi:10.1111/j.1468-1331.2011.03501.x
7. Shoesmith C, Abrahao A, Benstead T, et al. Canadian best practice recommendations for the management of amyotrophic lateral sclerosis. *CMAJ*. 2020;192(46):E1453-E1468. doi:10.1503/cmaj.191721.
8. ALS Association. El Escorial World Federation of Neurology criteria for the diagnosis of ALS. Available at: www.alsa.org/assets/pdfs/fyi/criteria_for_diagnosis-1.pdf.
9. ALS Functional Rating Scale. Available at: http://www.outcomes-umassmed.org/als/alsscale.aspx.

Effective date: 01/01/2025
Revised date: 11/14/2022