PHARMACY POLICY STATEMENT

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:

For _____ 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. 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

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For _____:

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

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