Sodium Phenylbutyrate/Taurursodiol (Relyvrio®)
Criteria for Use
November 2022

VA Pharmacy Benefits Management Services, Medical Advisory Panel, and VISN Pharmacist Executives

The following recommendations are based on medical evidence, clinician input, and expert opinion. The content of the document is dynamic and will be revised as new information becomes available. The purpose of this document is to assist practitioners in clinical decision-making, to standardize and improve the quality of patient care, and to promote cost-effective drug prescribing. THE CLINICIAN SHOULD USE THIS GUIDANCE AND INTERPRET IT IN THE CLINICAL CONTEXT OF THE INDIVIDUAL PATIENT. INDIVIDUAL CASES THAT ARE EXCEPTIONS TO THE EXCLUSION AND INCLUSION CRITERIA SHOULD BE ADJUDICATED AT THE LOCAL FACILITY ACCORDING TO THE POLICY AND PROCEDURES OF ITS P&T COMMITTEE AND PHARMACY SERVICES.

The Product Information should be consulted for detailed prescribing information.

See the VA National PBM-MAP-VPE Monograph on this drug at the *PBM INTERnet* or *PBM INTRAnet* site for further information.

Exclusion Criteria

If the answer to ANY item below is met, then the patient should **NOT** receive sodium phenylbutyrate/ taurursodiol.

- □ Notably decreased respiratory function and dyspnea (a total score of 3 or less points on ALSFRS-R^1 items for dyspnea, orthopnea, or respiratory insufficiency [the sum of questions 10, 11 and 12 on the ALSFRS-R])
- □ Bilevel positive airway pressure (BiPAP) dependent (use 24 hours per day)
- □ Other medical illness that will limit evaluation of motor function and efficacy (e.g. Parkinson's disease, schizophrenia, significant dementia, other major medical morbidity)
- □ Other possible causes for current symptoms as applicable not ruled out (e.g. cervical spondylosis or multifocal motor neuropathy)
- □ AST and/or ALT greater than 3 times upper limit of normal
- □ eGFR less than 60 mL/min/1.73m^2
- □ New York Heart Association (NYHA) class III/IV heart failure (due to high sodium content, 928 mg/day)

Inclusion Criteria

All the following must be fulfilled to receive sodium phenylbutyrate/taurursodiol.

- □ Care provided by a VA/ VA Community Care neurologist or locally designated ALS expert
- □ Diagnosis of ALS made or confirmed by a VA/VA Community Care neurologist or locally designated ALS expert
- □ Preserved function in either upper limbs (e.g. self-feed, self-dressing, typing) or lower limbs (e.g. walking)
- □ Discussion with the patient/caregiver/family regarding realistic treatment expectations and discontinuation of therapy should be documented in the patient's medical record.

[1] Amyotrophic Lateral Sclerosis Functional Rating Scale – Revised ALS-Functional-Rating-Scale-Revised-fill-in-form.pdf (encals.eu)

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