Generic Name: Sodium phenylbutyrate-taurursodiol

Applicable Drugs: Relyvrio™

Preferred: N/A

Non-preferred: N/A

Date of Origin: 2/27/2023

Date Last Reviewed / Revised: N/A

PRIOR AUTHORIZATION CRITERIA

(May be considered medically necessary when criteria I through IV are met)

I. Documented diagnosis of the following condition AND must meet criteria listed under applicable diagnosis:
   A. Amyotrophic lateral sclerosis (ALS) and BOTH criteria 1 and 2 are met:
      1. Documentation of slow vital capacity that is greater than 60% of predicted value for gender, height, and age.
      2. Patient is not dependent on invasive ventilation (i.e., no tracheostomy or tracheostomy for prevention of aspiration only).

II. Minimum age requirement: 18 years old.

III. Treatment must be prescribed by or in consultation with a neurologist.

EXCLUSION CRITERIA

- N/A

OTHER CRITERIA

- N/A

QUANTITY / DAYS SUPPLY RESTRICTIONS

- Quantities of up to 56 packets per 28 days.

APPROVAL LENGTH

- **Authorization:** 6 months
- **Re-Authorization:** An updated letter of medical necessity or progress notes showing positive clinical response (i.e., stabilization of ALS) AND absence of ventilator dependence (i.e., no tracheostomy or tracheostomy for prevention of aspiration only).

APPENDIX

N/A

REFERENCES

1. Relyvrio [package insert]. Amylyx Pharmaceuticals, Inc.; Cambridge, MA 02141; 2022.
2. Johnson SA, Fang T, De Marchi F, et al. Pharmacotherapy for amyotrophic lateral sclerosis: a review of approved and upcoming agents. *Drugs.* 2022;82(13):1367-1388. doi:10.1007/s40265-022-01769-1
3. Sun Y, Li X, Bedlack R. An evaluation of the combination of sodium phenylbutyrate and taurursodiol for the treatment of amyotrophic lateral sclerosis. *Expert Rev Neurother.* 2023;1-7. doi:10.1080/14737175.2023.2174018
4. 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
5. 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
6. Food and Drug Administration. FDA briefing document for the September 7, 2022 meeting of the Peripheral and Central Nervous System Drugs Advisory Committee. Posted September 7, 2022. Accessed January 14, 2023. https://www.fda.gov/media/161378/download