

CLINICAL CRITERIA: Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

Initial Approval: 6 months. Please note: “YES” responses would be considered EXCLUDED for Spinraza®.

Concomitant use of Zolgensma® (onasemnogene abeparvovec-xioi) with Spinraza® is considered investigational and not covered.

Has member tried Zolgensma®? Yes No

Spinraza® will only be approved after Zolgensma® use if there is documentation of clinical failure of Zolgensma® and the member qualifies for Spinraza® based on our criteria.

Does member have?

1. Respiratory insufficiency, defined by the medical necessity for invasive or non-invasive ventilation for greater than 6 hours during a 24-hour period, at screening? Yes No

2. Medical necessity for a gastric feeding tube, where this route gives the majority of nutrients? Yes No

3. Hypoxemia (O2 saturation awake less than 96%, without ventilation support)? Yes No

4. Presence of an implanted shunt for the drainage of CSF or an implanted CNS catheter? Yes No

5. Medical disability (e.g., wasting or cachexia, severe anemia, etc.) that would interfere with the assessment of safety? Yes No

6. Severe contracture(s) or severe scoliosis on radiograph (Cobb angle >40 degrees) that will effect intrathecal infusion? Yes No

7. Ability to walk independently (defined as the ability to walk unaided)? Yes No

8. Ability to walk with assistance? Yes No

9. HFMSE score >54? Yes No

Patient must have a diagnosis of 5q spinal muscular atrophy confirmed by **one** of the following (**ORIGINAL GENETIC labs included**):

Homozygous deletion of the SMN1 gene,

OR

Dysfunctional mutation of the SMN1 gene,

OR

Compound conversion mutation

AND

Documentation of genetic testing confirming no more than 4 copies of SMN2 and Type 1 -3;

(Continued on next page)

AND

- Documentation of baseline Movement assessments with **one** of the following:
 - Motor function/milestone: _____/32,

OR

- Hammersmith Infant Neurologic Exam (HINE): _____/68,

OR

- Hammersmith Functional Motor Scale for SMA (HFMS) _____/66 (**score >54 EXCLUDED**)

AND

- Baseline assessment of **one** of the following:
 - Number of hospitalization in the last 12 months _____

OR

- Baseline assessment of **one** of the following:
 - Number of hospitalization in the last 12 months _____

OR

- Number of antibiotic therapies for respiratory infection in the last 12 months _____

OR

- Current respiratory function test (e.g., forced vital capacity (FVC)): _____

Continuation Therapy. To qualify, check below all that apply. To support each line checked, all documentation (lab results, diagnostics, and/or chart notes) must be provided or request may be denied.

- Continuation of treatment with nusinersen **beyond six (6) months** after initiation of therapy **and every six (6) months thereafter** is considered medically necessary for the treatment of spinal muscular atrophy (SMA) when individuals meet the following:
 - Member has shown an improvement or no decrease from baseline score [**a decline from the baseline (6 months) over a 12-month evaluation would be considered not medically necessary**]; all three (3) assessments below would be reviewed from previous baseline:
 - Number of **hospitalization** in the **last 6 months**: _____
 - Number of **antibiotics** therapy for respiratory infection in the **last 6 months**: _____
 - Current respiratory function test [e.g., forced vital capacity (FVC)]: _____

AND

(Continued on next page)

- Documentation of Movement ASSESSMENT **within 30 days of request** must be provided or request may be denied:

- Motor function/milestone: _____/32;

OR

- Hammersmith Infant Neurologic Exam (HINE): _____/68;

OR

- Hammersmith Functional Motor Scale for SMA (HFMS): _____/66

AND

- Permanent ventilation defined as tracheostomy or ≥ 16 hours ventilator support per day would be considered a failure of Spinraza™ and will not be approved for continuation. Yes No

For urgent reviews: Practitioner should call Sentara Health Plans Pre-Authorization Department if they believe a standard review would subject the member to adverse health consequences. Sentara Health Plan's definition of urgent is a lack of treatment that could seriously jeopardize the life or health of the member or the member's ability to regain maximum function.

*****Use of samples to initiate therapy does not meet step edit/ preauthorization criteria.*****

****Previous therapies will be verified through pharmacy paid claims or submitted chart notes.****