

## nusinersen (Spinraza)

### Commercial Medical Benefit Drug Policy

#### Place of Service

Hospital Administration  
Office Administration  
Outpatient Facility Infusion Administration  
Infusion Center Administration  
Home Infusion

#### **Drug Details**

**USP Category:** GENETIC OR ENZYME OR PROTEIN DISORDER: REPLACEMENT, MODIFIERS, TREATMENT

**Mechanism of Action:** a survival motor neuron-2 (SMN2)-directed antisense oligonucleotide

#### **HCPCS:**

J2326:Injection, nusinersen, 0.1 mg

#### **How Supplied:**

12 mg (single-dose vial)

#### **Condition(s) listed in policy** *(see coverage criteria for details)*

- Spinal Muscular Atrophy (SMA)

Any condition not listed in this policy requires a review to confirm it is medically necessary. For conditions that have not been approved for intended use by the Food and Drug Administration (i.e., off-label use), the criteria outlined in the Health and Safety Code section 1367.21 must be met.

#### **Special Instructions and Pertinent Information**

Provider must submit documentation (such as office chart notes, lab results or other clinical information) to ensure the member has met all medical necessity requirements.

The member's specific benefit may impact drug coverage. Other utilization management processes, and/or legal restrictions may take precedence over the application of this clinical criteria.

For billing purposes, drugs must be submitted with the drug's assigned HCPCS code (as listed in the drug policy) and the corresponding NDC (national drug code). An unlisted, unspecified, or miscellaneous code should not be used if there is a specific code assigned to the drug.

#### **Coverage Criteria**

**The following condition(s) require Prior Authorization/Preservice.**

#### **Spinal Muscular Atrophy (SMA)**

**Meets medical necessity if all the following are met:**

#### Initial

1. Prescribed by or in consultation with a neurologist
2. Diagnosis of SMA confirmed by genetic testing demonstrating bi-allelic mutations in the survival motor neuron 1 (SMN1) gene by one of the following (a or b):
  - a. Deletion of both copies of the SMN1 gene
  - b. Identification of pathogenic variant(s) in both copies of the SMN1 gene
3. Genetic documentation of 4 or fewer copies of SMN2

4. Either of the following:
  - a. Provider attestation that patient has not received prior therapy with Zolgensma
  - b. Provider attestation that patient had an inadequate response with Zolgensma
5. Not being used in combination with Evrysdi or Zolgensma
6. **Effective 5/1/2026 and after:** Not being used in combination with Itvisma
7. Dose does not exceed the FDA-approved maximum

#### Reauthorization

1. Patient is responding based upon documentation of stabilization or improvement of continued motor function relative to projected natural course of SMA
2. **Effective 5/1/2026 and after:** Not being used in combination with Evrysdi, Zolgensma, or Itvisma
3. Dose does not exceed the FDA-approved maximum

#### **Covered Doses:**

Initial: Not to exceed 1 vial (12 mg) administered intrathecally every 2 weeks for 3 doses, followed by 1 vial one month after the 3rd dose; then 1 vial every 4 months thereafter

Maintenance: Not to exceed 1 vial (12 mg) administered intrathecally every 4 months

#### **Coverage Period:**

One year

#### **ICD-10:**

G12.0, G12.1

#### **References**

1. AHFS®. Available by subscription at <http://www.lexi.com>
2. DrugDex®. Available by subscription at <http://www.micromedexsolutions.com/home/dispatch>
3. Finkel RS, Mercuri E, Meyer OH, et al. Diagnosis and management of spinal muscular atrophy: Part 2: Pulmonary and acute care; medications, supplements and immunizations; other organ systems; and ethics. *Neuromuscul Disord NMD*. 2018;28(3):197-207.
4. Glascock J, Sampson J, Connolly AM, et al. Revised recommendations for the treatment of infants diagnosed with spinal muscular atrophy via newborn screening who have 4 copies of SMN2. *J Neuromuscul Dis* 2020; 7:97-100.
5. Institute for Clinical and Economic Review (ICER), Spinraza® and Zolgensma® for Spinal Muscular Atrophy: Effectiveness and Value. Final Evidence Report. April 3, 2019 (Updated May 24, 2019). Available from: [https://icer-review.org/wp-content/uploads/2018/07/ICER\\_SMA\\_Final\\_Evidence\\_Report\\_052419.pdf](https://icer-review.org/wp-content/uploads/2018/07/ICER_SMA_Final_Evidence_Report_052419.pdf). Accessed: May 24, 2019.
6. Mercuri, E, Bertini, E, Iannaccone, S. Childhood spinal muscular atrophy: controversies and challenges. *Lancet Neurol* 2012; 11(5): 443-452.
7. Mercuri E, Finkel R, Kirschner J et al. Efficacy and safety of nusinersen in children with later-onset spinal muscular atrophy (SMA): end of study results from the phase 3 CHERISH study. *Neuromuscular Disord* 2017; 27(Suppl 2):S210.

8. Mercuri E, Finkel RS, Muntoni F, et al. Diagnosis and management of spinal muscular atrophy: Part 1: Recommendations for diagnosis, rehabilitation, orthopedic and nutritional care. *Neuromuscul Disord NMD*. 2018;28(2):103-115.
9. Michelson D, Ciafaloni E, Ashwal S, et al. Evidence in focus: Nusinersen use in spinal muscular atrophy: Report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology. *Neurology*. 2018;91(20):923-933.
10. Spinraza (nusinersen) [prescribing information]. Cambridge, MA: Biogen; April 2024.
11. Wang CH, Finkel RS, Bertini ES, et al. Consensus statement for standard of care in spinal muscular atrophy. *J Child Neurol* 2007; 22:1027-1049. Available at: <http://journals.sagepub.com/doi/pdf/10.1177/0883073807305788>. Accessed on January 25, 2017.

### Review History

Date of Last Annual Review: 4Q2025

Changes from previous policy version:

- Spinal muscular atrophy:
  - ***Effective 5/1/2026 and after***, will manage combination use with Itvisma (Rationale: STEER trial)
  - ***Effective 5/1/2026 and after***, on reauthorization, will manage for combination with Evrysdi, Zolgensma, or Itvisma

*Blue Shield of California Medication Policy to Determine Medical Necessity  
Reviewed by P&T Committee*