Nusinersen (Spinraza®)	Place of Service Hospital Administration Office Administration Outpatient Facility Infusion Administration Infusion Center Administration Home Infusion HCPCS: **J2326** per 0.1 mg

Condition listed in policy *(see criteria for details)*

- Spinal muscular atrophy (SMA)

AHFS therapeutic class: Genetic disorder treatment; exon inclusion antisense oligonucleotide

Mechanism of action: antisense oligonucleotide shown in *in vitro* assays and studies in transgenic animal models of SMA to increase exon 7 inclusion in SMN2 messenger ribonucleic acid (mRNA) transcripts and production of full-length SMN protein.

(1) Special Instructions and Pertinent Information

Covered under the Medical Benefit, please submit clinical information for prior authorization review via fax.

(2) Prior Authorization/Medical Review is required for the following condition(s)

All requests for Spinraza® (nusinersen) must be sent for clinical review and receive authorization prior to drug administration or claim payment.

Spinal muscular atrophy (SMA)

1. Prescribed by or in consultation with a neurologist, **AND**
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. Deletion of both copies of the SMN1 gene, OR
    b. Identification of pathogenic variant(s) in both copies of the SMN1 gene

    AND
3. One of the following:
    a. Genetic documentation of 2 or 3 copies of SMN2, OR
    b. Genetic documentation of 4 copies of SMN2, and documentation of SMA-associated symptoms before 18 years of age

    AND
4. Either of the following:
    a. Provider attestation that patient has not received prior therapy with Zolgensma, or
    b. Provider attestation that patient had an inadequate response with Zolgensma,

    AND
5. Not being used in combination with Evrysdi

Covered Dose

- Initial: 1 vial (12 mg) administered intrathecally every 2 weeks x 3; followed by 1 vial one month later; then 1 vial every 4 months thereafter
- Maintenance: 1 vial every 4 months

Coverage Period

- Initial: 1 year
- Subsequent reauthorizations: Yearly based upon documentation of stabilization or improvement of continued motor function relative to projected natural course of SMA

ICD-10:
G12.0, G12.1

(3) The following condition(s) DO NOT require Prior Authorization/Preservice

All requests for Spinraza® (nusinersen) must be sent for clinical review and receive authorization prior to drug administration or claim payment.

(4) This Medication is NOT medically necessary for the following condition(s)

Blue Shield's research indicates there is inadequate clinical evidence to support off-label use of this drug for the following conditions (Health and Safety Code 1367.21):

- Type 4 SMA, also known as adult-onset SMA

Coverage for a Non-FDA approved indication, requires that criteria outlined in Health and Safety Code § 1367.21, including objective evidence of efficacy and safety are met for the proposed indication.

Please refer to the Provider Manual and User Guide for more information.

(5) Additional Information

How Supplied:

- 12 mg (single-dose vial)

(6) References

- AHFS®. Available by subscription at http://www.lexi.com
- DrugDex®. Available by subscription at http://www.micromedexsolutions.com/home/dispatch
- 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. doi:10.1016/j.nmd.2017.11.004
- 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. Accessed: May 24, 2019.
- Mercuri, E, Bertini, E, Iannaccone, S. Childhood spinal muscular atrophy: controversies and challenges. Lancet Neurol 2012; 11(5): 443-452.
- 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.
- 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.
- 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.
- Oskoui M, Levy G, Garland CJ et al. The changing natural history of spinal muscular atrophy type 1. Neurology 2007; 69: 1931-36.
- Spinraza® (nusinersen) [Prescribing information]. Cambridge, MA: Biogen Inc., 2/2023.
- 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.

(7) Policy Update

Date of last review: 4Q2023

Date of next review: 4Q2024

Changes from previous policy version:

- No clinical change to policy following routine annual review.

BSC Drug Coverage Criteria to Determine Medical Necessity
Reviewed by P&T Committee