

Spinraza® (nusinersen) (Intrathecal)

Document Number: IC-0291

Last Review Date: 08/08/2023

Date of Origin: 01/31/2017

Dates Reviewed: 01/2017, 02/2017, 01/2018, 08/2018, 06/2019, 08/2020, 08/2021, 08/2022, 08/2023

I. Length of Authorization

Coverage will be provided annually and may be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

- Loading: 1 vial on day 1, day 15, day 29, and day 59
- Maintenance: 1 vial every 112 days

B. Max Units (per dose and over time) [HCPCS Unit]:

- Loading: 120 billable units (12 mg) on day 1, day 15, day 29, and day 59
- Maintenance: 120 billable units (12 mg) every 112 days

III. Initial Approval Criteria

Submission of medical records (chart notes) related to the medical necessity criteria is **REQUIRED** on all requests for authorizations. Records will be reviewed at the time of submission. Please provide documentation related to diagnosis, step therapy, and clinical markers (i.e. genetic and mutational testing) supporting initiation when applicable. Please provide documentation via direct upload through the PA web portal or by fax.

Coverage is provided in the following conditions:

Universal Criteria ^{1,4,5,8}

- Patient must not have previously received treatment with SMA gene therapy (i.e., onasemnogene abeparvovec-xioi); **AND**
- Patient will not use in combination with other agents for SMA (e.g., onasemnogene abeparvovec-xioi, risdiplam, etc.); **AND**
- Patient must not have advanced disease (complete limb paralysis, permanent ventilation support, etc.); **AND**

- Patient must have the following laboratory tests at baseline and prior to each administration*: platelet count, prothrombin time, activated partial thromboplastin time, and quantitative spot urine protein testing; **AND**

Spinal Muscular Atrophy (SMA) † Φ 1-13

- Patient retains meaningful voluntary motor function (e.g., manipulate objects using upper extremities, ambulate, etc.); **AND**
- Patient must have a diagnosis of 5q spinal muscular atrophy confirmed by either homozygous deletion of the *SMN1* gene or dysfunctional mutation of the *SMN1* gene; **AND**
- Patient must have a diagnosis of SMA phenotype I, II, or III; **AND**
 - Patient has ≤ 3 copies of the *SMN2* gene (*Note: Patients with >3 copies of the SMN2 gene will be reviewed on a case-by-case basis*); **OR**
 - Patient has symptomatic disease (i.e., impaired motor function and/or delayed motor milestones); **AND**
- Baseline documentation of one or more of the following:
 - Motor function/milestones, including but not limited to, the following validated scales: Hammersmith Infant Neurologic Exam (HINE), Hammersmith Functional Motor Scale Expanded (HF MSE), Children’s Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND), Bayley Scales of Infant and Toddler development Third Ed. (BSID-III), 6-minute walk test (6MWT), Upper Limb Module (ULM), Motor Function Measure 32 (MFM32), Revised Upper Limb Module (RULM), etc.
 - Respiratory function tests [e.g., forced vital capacity (FVC), etc.]
 - Exacerbations necessitating hospitalization and/or antibiotic therapy for respiratory infection in the preceding year/timeframe
 - Patient weight (for patients without a gastrostomy tube)

*Laboratory tests should be obtained within several days prior to administration

† FDA Approved Indication(s), ‡ Compendia Recommended Indication(s); Φ Orphan Drug

IV. Renewal Criteria 1-13

Coverage can be renewed based upon the following criteria:

- Patient continues to meet the universal and other indication-specific relevant criteria such as concomitant therapy requirements (not including prerequisite therapy), performance status, etc. identified in section III; **AND**
- Absence of unacceptable toxicity which would preclude safe administration of the drug. Examples of unacceptable toxicity include: significant renal toxicity, thrombocytopenia, coagulation abnormalities, etc.; **AND**

- Patient has responded to therapy compared to pretreatment baseline in one or more of the following:
 - Stability or improvement in net motor function/milestones, including but not limited to, the following validated scales: Hammersmith Infant Neurologic Exam (HINE), Hammersmith Functional Motor Scale Expanded (HFMSE), Children’s Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND), Bayley Scales of Infant and Toddler development Third Ed. (BSID-III), 6-minute walk test (6MWT), Upper Limb Module (ULM), Motor Function Measure 32 (MFM32), Revised Upper Limb Module (RULM), etc.
 - Stability or improvement in respiratory function tests [e.g., forced vital capacity (FVC), etc.]
 - Reduction in exacerbations necessitating hospitalization and/or antibiotic therapy for respiratory infection in the preceding year/timeframe
 - Stable or increased patient weight (for patients without a gastrostomy tube)
 - Slowed rate of decline in the aforementioned measures

V. Dosage/Administration ¹

Indication	Dose
Spinal Muscular Atrophy	<p>Administer 12 mg as an intrathecal bolus injection over 1 to 3 minutes using a spinal anesthesia needle. Prior to administration, 5 mL of cerebrospinal fluid should be removed. Imaging guidance and sedation may be required for administration.</p> <p><u>Initiation</u></p> <p>Four loading doses: The first three loading doses should be administered at 14-day intervals. The 4th loading dose should be administered 30 days after the 3rd dose.</p> <p><u>Maintenance</u></p> <p>One dose every 4 months (112 days) thereafter</p>
NOTE:	
<ul style="list-style-type: none"> • Store refrigerated at 2°C to 8°C (36°F to 46°F) in the original carton to protect from light. Do not freeze. • If no refrigeration is available, Spinraza may be stored in its original carton, protected from light at or below 30°C (86°F) for up to 14 days. • Allow Spinraza to warm to room temperature (25°C / 77°F) prior to administration. 	

VI. Billing Code/Availability Information

HCPCS code:

- J2326 –Injection, nusinersen, 0.1 mg; 1 billable unit = 0.1 mg

NDC:

- Spinraza 12 mg/5 mL solution for injection; single-dose vial: 64406-0058-xx

VII. References

1. Spinraza [package insert]. Cambridge, MA; Biogen, Inc.; February 2023. Accessed July 2023.
2. Wang CH, Finkel RS, Bertini ES, et al. Consensus statement for standard of care in spinal muscular atrophy. *J Child Neurol.* 2007 Aug;22(8):1027-49.
3. Prior TW, Finanger E. Spinal muscular atrophy. GeneReviews. www.ncbi.nlm.nih.gov/books/NBK1352/. Initial Posting: February 24, 2000; Last Revision: December 3, 2022. Accessed on July 12, 2023.
4. Finkel RS, Mercuri E, Darras BT, et al; for the ENDEAR Study Group. Nusinersen versus sham control in infantile-onset spinal muscular atrophy. *N Engl J Med.* 2017;377(18):1723-1732. Finkel RS, Mercuri E, Darras BT, et al; for the ENDEAR Study Group. Nusinersen versus sham control in infantile-onset spinal muscular atrophy. *N Engl J Med.* 2017;377(18):1723-1732.
5. Mercuri E, Darras BT, Chiriboga CA, et al; for the CHERISH Study Group. Nusinersen versus sham control in later-onset spinal muscular atrophy. *N Engl J Med.* 2018 Feb 15;378(7):625-635. doi: 10.1056/NEJMoa1710504.
6. Dabbous O, Maru B, Jansen JP, et al. Survival, Motor Function, and Motor Milestones: Comparison of AVXS-101 Relative to Nusinersen for the Treatment of Infants with Spinal Muscular Atrophy Type 1. *Adv Ther.* 2019 May;36(5):1164-1176.
7. Kichula E, Duong T, Glanzman A, et al. Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND) Feasibility for Individuals with Severe Spinal Muscular Atrophy II (S46.004). *Neurology* Apr 2018, 90 (15 Supplement) S46.004
8. De Vivo DC, Bertini E, Swoboda KJ, et al. Nusinersen initiated in infants during the presymptomatic stage of spinal muscular atrophy: Interim efficacy and safety results from the Phase 2 NURTURE study. *Neuromuscul Disord.* 2019 Nov;29(11):842-856. Doi: 10.1016/j.nmd.2019.09.007. Epub 2019 Sep 12.
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 Nov 13;91(20):923-933. Doi: 10.1212/WNL.0000000000006502. Epub 2018 Oct 12.
10. Darras BT, Chiriboga CA, Iannaccone ST, et al. Nusinersen in later-onset spinal muscular atrophy: long-term results from the phase 1/2 studies. *Neurology.* 2019;92(21):e2492-e2506
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12. (ICER) IfCaER . Spinraza and Zolgensma for Spinal Muscular Atrophy: Effectiveness and Value. Final Evidence Report. April 3, 2019 (Updated May 24, 2019) 2019.
13. Kichula E, Duong T, Glanzman A, et al. Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND) Feasibility for Individuals with Severe Spinal Muscular Atrophy II (S46.004). *Neurology* Apr 2018, 90 (15 Supplement) S46.004

14. Noridian Healthcare Solutions, LLC. Local Coverage Article: Billing and Coding: Spinraza® (nusinersen) (A58578). Centers for Medicare & Medicaid Services, Inc. Updated on 11/18/2021 with effective date of 11/25/2021. Accessed July 2023.
15. Noridian Healthcare Solutions, LLC. Local Coverage Article: Billing and Coding: Spinraza® (nusinersen) (A58579). Centers for Medicare & Medicaid Services, Inc. Updated on 11/18/2021 with effective date of 11/25/2021. Accessed July 2023.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
G12.0	Infantile spinal muscular atrophy, type I [Werdnig-Hoffmann]
G12.1	Other inherited spinal muscular atrophy
G12.25	Progressive spinal muscle atrophy
G12.8	Other spinal muscular atrophies and related syndromes
G12.9	Spinal muscular atrophy, unspecified

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: <http://www.cms.gov/medicare-coverage-database/search.aspx>. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD/LCA):

Jurisdiction(s): E	NCD/LCD/LCA Document (s): A58578
https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a58578&areaId=all&docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP	

Jurisdiction(s): F	NCD/LCD/LCA Document (s): A58579
https://www.cms.gov/medicare-coverage-database/new-search/search-results.aspx?keyword=a58579&areaId=all&docType=NCA%2CCAL%2CNCD%2CMEDCAC%2CTA%2CMCD%2C6%2C3%2C5%2C1%2CF%2CP	

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	Applicable State/US Territory	Contractor
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC

Medicare Part B Administrative Contractor (MAC) Jurisdictions

Jurisdiction	Applicable State/US Territory	Contractor
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC