Spinraza® (nusinersen)
(Intrathecal)

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I. Length of Authorization

- Initial: Prior authorization validity will be provided initially for 12 months (365 days).
- Renewal: Prior authorization validity may be renewed every 12 months (365 days) thereafter.

II. Dosing Limits

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

- Loading: 500 billable units (50 mg) on days 1,15, 29, & 59
- Maintenance: 280 billable units (28 mg) every 112 days

III. Initial Approval Criteria

Submission of supporting clinical documentation (including but not limited to medical records, chart notes, lab results, and confirmatory diagnostics) related to the medical necessity criteria is REQUIRED on all requests for authorizations. Records will be reviewed at the time of submission as part of the evaluation of this request. 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. Failure to submit the medical records may result in the denial of the request due to inability to establish medical necessity in accordance with policy guidelines.

Prior authorization validity is provided in the following conditions:

Universal Criteria [1,4,5,8]

- Member will not use in combination with other agents for Spinal Muscular Atrophy (SMA) (e.g., onasemnogene abeparvovec-xioi, risdiplam, etc.); **AND**
- Member must not have advanced disease (e.g., complete limb paralysis, permanent ventilation support, etc.); **AND**
- Member 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**

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

Spinal Muscular Atrophy (SMA) † Φ [1-15]

- Member retains meaningful voluntary motor function (e.g., manipulate objects using upper extremities, ambulate, etc.); **AND**
- Member must have a diagnosis of 5q spinal muscular atrophy confirmed by either homozygous deletion or dysfunctional mutation of the *SMN1* gene; **AND**
- Member must have a diagnosis of SMA phenotype 1, 2, or 3; **AND**
  - Member has ≤ 3 copies of the *SMN2* gene *(Note: Members with >3 copies of the SMN2 gene will be reviewed on a case-by-case basis)*; **OR**
  - Member 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 (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.
  - 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
  - Member weight (for members without a gastrostomy tube)

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

IV. Renewal Criteria [1-13]

Prior authorization validity may be renewed based upon the following criteria:

- Member 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**
  - Member 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 member weight (for members without a gastrostomy tube)
- Slowed rate of decline in the aforementioned measures; **OR**

- Member has initially responded to therapy with a subsequent loss of response and/or increase in weakness (e.g., wearing off effect) prior to the next scheduled dose of the Low Dose Regimen and warrants an increase in dose to the High Dose Regimen; **AND**
  - Member is receiving a Low-Dose Regimen and has received all loading doses and at least one maintenance dose

V. Dosage/Administration [1]

<table>
<tr><th>Indication</th><th>Dose</th></tr>
<tr><td>Spinal Muscular Atrophy</td><td>
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.
<table>
<tr><th></th><th>Loading Dosages</th><th>Maintenance Dosage</th></tr>
<tr><td>Low Dose Regimen
(Low dose with 4 loading doses)</td><td>Administer a total of 4 loading doses as follows:
one 12 mg dose every 14 days for three doses, then
a fourth 12 mg dose 30 days after the third dose</td><td>Administer 12 mg once every 4 months starting 4 months after the last loading dose</td></tr>
<tr><td>High Dose Regimen
(High dose with 2 loading doses)</td><td>Administer a total of 2 loading doses as follows:
one 50 mg dose followed by a second 50 mg dose 14 days later</td><td>Administer 28 mg once every 4 months starting 4 months after the last loading dose</td></tr>
</table>
<u>Transitioning Between the Low Dose and High Dose Regimens</u>
If transitioning from SPINRAZA Low Dose Regimen to High Dose Regimen, administer a single 50 mg bolus dose at least four months (+/- 14 days) after the last 12 mg maintenance dose, followed by a 28 mg maintenance dose once every 4 months thereafter.
</td></tr>
<tr><td colspan="2"><u>NOTE:</u>
Store refrigerated at 2ºC to 8ºC (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 30ºC (86ºF) for up to 14 days.</td></tr>
</table>

- Allow the Spinraza vial to warm to room temperature (25ºC / 77ºF) 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
- Spinraza 28 mg/5 mL solution for injection; single-dose vial: 64406-0036-xx
- Spinraza 50 mg/5 mL solution for injection; single-dose vial: 64406-0037-xx

VII. References

1. Spinraza [package insert]. Cambridge, MA; Biogen; March 2026. Accessed March 2026.
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Appendix A – Non-Quantitative Treatment Limitations (NQTL) Factor Checklist

Non-quantitative treatment limitations (NQTLs) refer to the methods, guidelines, standards of evidence, or other conditions that can restrict how long or to what extent benefits are provided under a health plan. These may include things like utilization review or prior authorization. The utilization management NQTL applies comparably, and not more stringently, to mental health/substance use disorder (MH/SUD) Medical Benefit Prescription Drugs and medical/surgical (M/S) Medical Benefit Prescription Drugs. The table below lists the factors that were considered in designing and applying prior authorization to this drug/drug group, and a summary of the conclusions that Prime's assessment led to for each.

Factor	Conclusion
Indication	Yes: Consider for PA
Safety and efficacy	No: PA not a priority
Potential for misuse/abuse	No: PA not a priority
Cost of drug	Yes: Consider for PA

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

ICD-10	ICD-10 Description
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)

The preceding information is intended for non-Medicare coverage determinations. 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 Determinations (NCDs) and/or Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD, or LCA documents: https://www.cms.gov/medicare-coverage-database/search.aspx. Additional indications, including any preceding information, may be applied at the discretion of the health plan.

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

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdictio	**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
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
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA
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