



Spinraza™ (nusinersen) (Intrathecal)

Document Number: IC-0291

Last Review Date: 08/02/2018

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Dates Reviewed: 01/2017, 02/2017, 01/2018, 08/2018

I. Length of Authorization

Coverage will be provided annually and may be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [Pharmacy Benefit]:

- Loading: 1 vial on D1, D15, D29, and D59
- Maintenance: 1 vial every 112 days

B. Max Units (per dose and over time) [Medical Benefit]:

- Loading: 120 billable units on D1, D15, D29, and D59
- Maintenance: 120 billable units every 112 days

III. Initial Approval Criteria

Coverage is provided in the following conditions:

Spinal Muscular Atrophy (SMA) †

- 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**
- 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 one of the following SMA phenotypes; **AND**
 - SMA I

- SMA II with symptomatic disease (i.e. impaired motor function and/or delayed motor milestones)
- SMA III with symptomatic disease (i.e. impaired motor function and/or delayed motor milestones)
- 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 (HFMSSE), 6-minute walk test (6MWT), upper limb module (ULM), 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)

† FDA-labeled indication(s)

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

IV. Renewal Criteria

- Patient continues to meet the criteria in Section III; **AND**
- Absence of unacceptable toxicity which would preclude safe administration of the drug. Examples of unacceptable toxicity include the following: 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 (HFMSSE), 6-minute walk test (6MWT), upper limb module (ULM), 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

Indication	Dose
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Spinal Muscular Atrophy	12 mg administered, as an intrathecal bolus injection over 1 to 3 minutes using a spinal anesthesia needle, per administration. Prior to administration, 5 mL of cerebrospinal fluid should be removed. Imaging guidance and sedation may be required for administration.
	<u>Initiation</u>
	Four loading doses: the first three loading doses should be administered at 14-day intervals. The 4 th loading dose should be administered 30 days after the 3 rd dose.
	<u>Maintenance</u>
	One dose every 112 days thereafter

Store refrigerated at 2°C to 8°C; warm to room temperature prior to administration

VI. Billing Code/Availability Information

Jcode:

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.; May 2018. Accessed June 2018.
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/ (Accessed on December 12, 2017).
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;375:625-635.

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/advanced-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): N/A

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
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 Government Benefit Administrators, 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