# Pharmacy Medical Necessity Guidelines: Spinraza (nusinersen)

*Effective: April 1, 2021*

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|---|---|---|---|
| Prior Authorization Required | √ | Type of Review – Care Management | |
| Not Covered | | Type of Review – Clinical Review | √ |
| Pharmacy (RX) or Medical (MED) Benefit | MED | Department to Review | PRECERT/ MM |

These pharmacy medical necessity guidelines apply to the following:

**Commercial Products**

☒ Tufts Health Plan Commercial products – large group plans
☒ Tufts Health Plan Commercial products – small group and individual plans
☒ Tufts Health Freedom Plan products – large group plans
☒ Tufts Health Freedom Plan products – small group plans
- CareLink℠ – Refer to CareLink Procedures, Services and Items Requiring Prior Authorization

**Tufts Health Public Plans Products**

☒ Tufts Health Direct – A Massachusetts Qualified Health Plan (QHP) (a commercial product)
☒ Tufts Health Together – MassHealth MCO Plan and Accountable Care Partnership Plans
☒ Tufts Health RITogether – A Rhode Island Medicaid Plan

**Fax Numbers:**

Commercial Products:
PRECERT: 617.972.9409

Tufts Health Public Plans Products:
MM: 888.415.9055

**Note:** This guideline does not apply to Medicare Members (includes dual eligible Members).

## OVERVIEW

**FOOD AND DRUG ADMINISTRATION-APPROVED INDICATION(S)**

Spinraza (nusinersen) is a survival motor neuron-2-directed antisense oligonucleotide indicated for the treatment of spinal muscular atrophy (SMA) in pediatric and adult patients.

## COVERAGE GUIDELINES

The plan may authorize coverage of Spinraza (nusinersen) for Members, when all of the following criteria are met:

**Initial Authorization**

1. Documented diagnosis of spinal muscular atrophy type 1, 2, or 3 confirmed by molecular genetic testing of any of the following: SMN1 homozygous gene deletion, homozygous conversion mutation (i.e., SMN1 gene conversion to SMN2 gene), or compound heterozygote mutation

**AND**

2. Prescribed by or written in consultation with a board-certified neurologist with special qualification in child neurology and treatment of spinal muscular atrophy

**AND**

3. Documentation of baseline (pre-treatment) motor function skills

**AND**

4. The Member is not using Spinraza concomitantly with Evrysdi

**AND**

5. Documentation of one of the following:
   a. The Member has not previously received gene replacement therapy for the treatment of SMA
   b. The Member has previously received gene replacement therapy for the treatment of SMA and has experienced a decline in clinical status since receipt of gene replacement therapy

**AND**

6. Documentation the Member is not ventilation dependent (defined as using a ventilator 16 hours or more a day)

**Reauthorization Criteria**

1. Documented diagnosis of spinal muscular atrophy type 1, 2, or 3 confirmed by molecular genetic testing

**AND**

2. Prescribed by or in written in consultation with a board-certified neurologist with special qualification in child neurology and treatment of spinal muscular atrophy

**AND**

3. The Member is not using Spinraza concomitantly with Evrysdi

**AND**

4. Documentation of one of the following:
    a. The Member has not previously received gene replacement therapy for the treatment of spinal muscular atrophy
    b. The Member has previously received gene replacement therapy for the treatment of spinal muscular atrophy and has experienced a decline in clinical status since receipt of gene replacement therapy

**AND**

5. Documentation the Member is not ventilation dependent (defined as using a ventilator 16 hours or more a day)

**AND**

6. Documentation of disease stabilization or clinical improvement of SMA symptoms (e.g., limb and trunk weakness; hypotonia and impaired head control; difficulty breathing, swallowing, feeding, and handling secretions

## LIMITATIONS

- Coverage of Spinraza (nusinersen) for SMA type 4 will not be authorized.
- Authorizations will be provided for 12 months.
- Members new to the plan stable on Spinraza (nusinersen) should be reviewed against Initial Authorization Criteria. For treatment-experienced Members, Providers must submit documentation of a physical assessment, motor function-based testing, and need for medical intervention related to SMA symptoms, relative to baseline.
- If gene therapy is subsequently administered, Spinraza authorization will be terminated and the Member must reapply for coverage.

## CODES

The following HCPCS/CPT code(s) are:

| Code | Description |
|---|---|
| J2326 | Injection, nusinersen, 0.1 mg |

## REFERENCES


1. Chiriboga CA, Swoboda KJ, Darras BT, et al. Results from a phase 1 study of nusinersen (ISIS-SMN(Rx)) in children with spinal muscular atrophy. Neurology. 2016;86(10):890-897
2. CureSMA. SPINRAZA (Nusinersen) URL: curesma.org/spinraza/. Available on Internet: Accessed 2018 May 17.
3. Finkel RS, Chiriboga CA, Vajsar J, et al. Treatment of infantile-onset spinal muscular atrophy with nusinersen: A phase 2, open-label, dose-escalation study. Lancet. 2017;388(10063):3017-3026.
4. Haché M, Swoboda KJ, Sethna N, et al. Intrathecal injections in children with spinal muscular atrophy: Nusinersen clinical trial experience. J Child Neurol. 2016;31(7):899-906.
5. 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*. 2018; 28(2): 103-15.
6. Mercuri E, Darras BT, Chiriboga CA, et al. Nusinersen versus sham control in later-onset spinal muscular atrophy. N Engl J Med. 2018;378:625-35.
7. Spinraza (nusinersen) [prescribing information]. Cambridge, MA: Biogen Inc; October 2018.
8. Wang CH, Finkel RS, Bertini ES, et al. Consensus statement for standard of care in spinal muscular atrophy. J Child Neurol. Aug 2007;22(8):1027-49.
9. Zolgensma (onasemnogene abeparvovec-xioi )[prescribing information]. Bannockburn, IL: AveXis, Inc; May 2019.


## APPROVAL HISTORY

June 13, 2017: Reviewed by Pharmacy & Therapeutics Committee.

Subsequent endorsement date(s) and changes made:

1. January 1, 2018: Administrative update: Added new J code J2326 to Medical Necessity Guideline and removed expired C code C9489.
2. June 12, 2018: No changes.
3. March 12, 2019: No changes.
4. July 9, 2019: Effective October 1, 2019, added criteria requiring documentation the Member has not been previously treated with gene therapy and the limitations stating coverage would not be authorized for members who have been previously treated with gene therapy (for example,

Zolgensma (onasemnogene abeparvovec-xioi)), and coverage would be terminated if gene therapy is administered, as the drug has not been studied in combination.

5. November 12, 2019: Effective April 1, 2020, moved reauthorization criteria from the Limitations section to the Coverage Guidelines section to ensure appropriate application of criteria.
6. December 8, 2020: No changes.
7. January 12, 2021: Effective April 1, 2021, updated criteria to require documentation of one of the following: "The Member has not previously received gene replacement therapy for the treatment of spinal muscular atrophy or the Member has previously received gene replacement therapy for the treatment of spinal muscular atrophy and has experienced a decline in clinical status since receipt of gene replacement therapy." Removed the following Limitation "Coverage of Spinraza (nusinersen) will not be authorized for Members who have been previously treated with gene therapy, for example Zolgensma (onasemnogene abeparvovec-xioi)." Updated provider specialty requirements to "Prescribed by or written in consultation with a board-certified neurologist with special qualification in child neurology and treatment of spinal muscular atrophy." Added the following requirements to the coverage criteria: "The Member is not using Spinraza concomitantly with Evrysdi" and "Documentation the Member is not ventilation dependent (defined as using a ventilator 16 hours or more a day)." Updated the Limitations language to clarify that Members new to the plan stable on Spinraza (nusinersen) should be reviewed against Initial Authorization Criteria.

## BACKGROUND, PRODUCT AND DISCLAIMER INFORMATION

Pharmacy Medical Necessity Guidelines have been developed for determining coverage for plan benefits and are published to provide a better understanding of the basis upon which coverage decisions are made. The plan makes coverage decisions on a case-by-case basis considering the individual member's health care needs. Pharmacy Medical Necessity Guidelines are developed for selected therapeutic classes or drugs found to be safe, but proven to be effective in a limited, defined population of patients or clinical circumstances. They include concise clinical coverage criteria based on current literature review, consultation with practicing physicians in the service area who are medical experts in the particular field, FDA and other government agency policies, and standards adopted by national accreditation organizations. The plan revises and updates Pharmacy Medical Necessity Guidelines annually, or more frequently if new evidence becomes available that suggests needed revisions.

For self-insured plans, coverage may vary depending on the terms of the benefit document. If a discrepancy exists between a Pharmacy Medical Necessity Guideline and a self-insured Member's benefit document, the provisions of the benefit document will govern.

Treating providers are solely responsible for the medical advice and treatment of members. The use of this policy is not a guarantee of payment or a final prediction of how specific claim(s) will be adjudicated. Claims payment is subject to member eligibility and benefits on the date of service, coordination of benefits, referral/authorization and utilization management guidelines when applicable, and adherence to plan policies and procedures and claims editing logic.