Harvard Pilgrim Health Care | TUFTS Health Plan

Medical Necessity Guidelines
Medical Benefit Drugs
Spinraza® (nusinersen)

Effective: January 1, 2025

Guideline Type	☒ Prior Authorization ☐ Non-Formulary ☐ Step-Therapy ☐ Administrative

Applies to:

Commercial Products

☒ Harvard Pilgrim Health Care Commercial products; Fax 617-673-0988
☒ Tufts Health Plan Commercial products; Fax 617-673-0988
CareLink℠ – Refer to CareLink Procedures, Services and Items Requiring Prior Authorization

Public Plans Products

☒ Tufts Health Direct – A Massachusetts Qualified Health Plan (QHP) (a commercial product); Fax 617-673-0988
☐ Tufts Health Together – MassHealth MCO Plan and Accountable Care Partnership Plans; Fax 617-673-0939
☒ Tufts Health RITogether – A Rhode Island Medicaid Plan; Fax 617-673-0939
☐ Tufts Health One Care* – A Medicare-Medicaid Plan (a dual eligible product); Fax 617-673-0956
*The MNG applies to Tufts Health One Care members unless a less restrictive LCD or NCD exists.

Senior Products

☐ Harvard Pilgrim Health Care Stride Medicare Advantage; Fax 617-673-0956
☐ Tufts Health Plan Senior Care Options (SCO), (a dual-eligible product); Fax 617-673-0956
☐ Tufts Medicare Preferred HMO, (a Medicare Advantage product); Fax 617-673-0956
☐ Tufts Medicare Preferred PPO, (a Medicare Advantage product); Fax 617-673-0956

Note: While you may not be the provider responsible for obtaining prior authorization, as a condition of payment you will need to ensure that prior authorization has been obtained.

Overview

Food and Drug Administration – Approved Indications

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

Clinical Guideline Coverage Criteria

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

Initial Authorization Criteria

1. Documented diagnosis of spinal muscular atrophy confirmed by molecular genetic testing of any of the following: SMN1 homozygous gene deletion, homozygous gene mutation, or compound heterozygous mutation

AND

2. Prescribed by or 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 as assessed by **one (1)** of the following:
   a. Hammersmith Infant Neurological Exam Part 2 (HINE-2)
   b. Hammersmith Functional Motor Scale Expanded (HFMSE)

c. Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND)

AND

4. The patient is not using Spinraza concomitantly with Evrysdi

AND

5. Documentation of **one (1)** of the following:
   a. The patient has not previously received gene replacement therapy for the treatment of spinal muscular atrophy
   b. Both of the following:
      i. The patient has previously received gene replacement therapy for the treatment of spinal muscular atrophy
      ii. The patient has experienced a decline in clinical status since receipt of gene replacement therapy

AND

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

Reauthorization Criteria

1. Documented diagnosis of spinal muscular atrophy confirmed by molecular genetic testing

AND

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

AND

3. The patient is not using Spinraza concomitantly with Evrysdi

AND

4. Documentation of **one (1)** of the following:
   a. The patient has not previously received gene replacement therapy for the treatment of spinal muscular atrophy
   c. Both of the following:
      i. The patient has previously received gene replacement therapy for the treatment of spinal muscular atrophy
      ii. The patient has experienced a decline in clinical status since receipt of gene replacement therapy

AND

5. Documentation the patient 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 spinal muscular atrophy symptoms as demonstrated by at least **one (1)** of the following assessments:
   a. **Both** of the following with respect to HINE-2
      a. **One** of the following:
         i. Patient exhibited improvement or maintenance of previous improvement of at least a 2-point (or maximal score) increase in ability to kick
         ii. Patient exhibited improvement or maintenance of previous improvement of at least a 1-point (or maximal score) increase in any other HINE-2 milestone (e.g., head control, rolling, sitting, crawling, standing, or walking) excluding voluntary grasp
      b. **One** of the following:
         i. Patient exhibited improvement or maintenance of previous improvement in more HINE-2 motor milestones than worsening (net positive improvement)
         ii. Patient achieved and maintained any new motor milestones when they would otherwise be unexpected to do so (e.g., sit or stand unassisted, walk)
   b. **One (1)** of the following with respect to HFMSE
      a. Patient exhibited improvement or maintenance of previous improvement of at least a 3-point increase in score
      b. Patient has achieved and maintained any new motor milestone from pretreatment baseline when they would otherwise be unexpected to do so
   c. **One (1)** of the following with respect to CHOP-INTEND
      a. Patient exhibited improvement or maintenance of previous improvement of at least a 4-point increase in score
      b. Patient has achieved and maintained any new motor milestone from pretreatment baseline when they would otherwise be unexpected to do so

Limitations

- Authorizations will be provided for 12 months.
- Members new to the plan stable on Spinraza 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 code(s) require prior authorization:

Table 1: HCPCS Codes

HCPCS Codes	Description
J2326	Injection, nusinersen, 0.1 mg

References

1. Calucho M, Bernal S, Alias L, et al. Correlation between SMA type and SMN2 copy number revisited: An analysis of 625 unrelated Spanish patients and a compilation of 2834 reported cases. Neuromuscul Disord. 2018;28(3):208-215
2. 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
3. CureSMA. SPINRAZA (Nusinersen) URL: curesma.org/spinraza/. Available on Internet: Accessed 2018 May 17.
4. 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.
5. 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.
6. 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.
7. Mercuri E, Darras BT, Chiriboga CA, et al. Nusinersen versus sha m control in later-onset spinal muscular atrophy. N Engl J Med. 2018;378:625-35.
8. SMA Overview. https://smafoundation.org/about-sma/materials/. Accessed January 6, 2022.
9. Spinraza (nusinersen) [prescribing information]. Cambridge, MA: Biogen Inc; April 2024.
10. Verhaart IEC, Robertson A, Wilson IJ, et al. Prevalence, incidence and carrier frequency of 5q-linked spinal muscular atrophy – a literature review. Orphanet J Rare Dis. 2017;12(1):124.
11. Wang C et al. Consensus Statement for Standard of Care in Spinal Muscular Atrophy. Journal of Child Neurology, Volume 22 Number 8. August 2007.
12. 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.

Approval And Revision History

September 13, 2022: Reviewed by Pharmacy and Therapeutics Committee (P&T)

Subsequent endorsement date(s) and changes made:

- September 21, 2022: Reviewed by the Medical Policy Approval Committee (MPAC)
- November 14, 2023: Removed the Limitation Coverage will be limited to the FDA-approved dosing regimen. Removed the Clinical classification for SMA (Table adapted from RS Finkel et al, 2017, p.597) (effective 12/1/2023).
- November 2023: Administrative update to rebrand Tufts Health Unify to Tufts Health One Care for 2024.
- November 2024: Effective 1/1/2025 - removed the specific exam/scoring requirements for the patients who have experienced a decline in clinical status since receipt of gene replacement therapy. This change was made to simplify criteria and is in line with criteria for drugs in this class (Evrysdi). Administrative change to the reference section.

Background, Product and Disclaimer Information

Medical Necessity Guidelines are developed to determine coverage for benefits and are published to provide a better understanding of the basis upon which coverage decisions are made. We make coverage decisions using these guidelines, along with the Member's benefit document, and in coordination with the Member's physician(s) on a case-by-case basis considering the individual Member's health care needs.

Medical Necessity Guidelines are developed for selected therapeutic or diagnostic services found to be safe and proven 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 our service area who are medical experts in the particular field, FDA and other government agency policies, and standards adopted by national accreditation organizations. We revise and update 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 Medical Necessity Guideline and a self-insured Member's benefit document, the provisions of the benefit document will govern. For Tufts Health Together (Medicaid), coverage may be available beyond these guidelines for pediatric members under age 21 under the Early and Periodic Screening, Diagnostic and Treatment (EPSDT) benefits of the plan in accordance with 130 CMR 450.140 and 130 CMR 447.000, and with prior authorization.

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