SmartHealth Ascension

Nusinersen

(Spinraza®) J2326 (0.1mg)

Covered with prior authorization

Nusinersen (Spinraza®) may be authorized when the following criteria are met:

- Diagnosis of **spinal muscular atrophy (SMA)** by neurologist or pediatric neuromuscular specialist; **AND**
- Diagnosis confirmed by
    - Spinal Muscular Atrophy (SMA) diagnostic test results confirming 0 copies of SMN1; **OR**
    - Molecular genetic testing of 5q SMA for any of the following:
        - homozygous gene deletion; **OR**
        - homozygous conversion mutation; **OR**
        - compound heterozygote; **AND**
- Documentation is provided that individual has either:
    - Genetic testing confirming no more than 2 copies of SMN2; **OR**
    - Onset of SMA-associated signs and symptoms before 21 months of age; **AND**
- Type 1, Type 2 with symptomatic disease, or Type 3 with symptomatic disease; **AND**
    - Individual is 15 years of age or younger at initiation of treatment; **AND**
    - Symptomatic individuals must be able to sit independently; **AND**
    - Individuals must not be dependent on either of the following**:**
        - Invasive ventilation or tracheostomy; **OR**
        - Use of non-invasive ventilation (BiPAP) for more than 16 hours per day; **AND**
- Documentation of baseline motor function by one of the following exams:
    - Hammersmith Infant Neurologic Exam [HINE] (infant to early childhood); **OR**
    - Hammersmith Functional Motor Scale Expanded [HFMSE]; **OR**
    - Upper Limb Module [ULM] test (non-ambulatory); **OR**
    - Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders [CHOP-INTEND]; **AND**
- Individual has not received ZOLGENSMA® (onasemnogene abeparvovec-xioi) or cell therapy; **UNLESS**
    - All other criteria are met; **AND**
    - Documentation is provided that individual has experienced a decline in clinical status since receiving gene therapy; **AND**

- Prescribed by a neurologist or pediatric neuromuscular; **AND**
- The prescribed regimen is within the FDA-approved dosing regimen:
    - Loading dose(s):
        - 12 mg once every 14 days for 3 doses; then,
        - 12 mg once 30 days after the third dose.
    - Maintenance: 12 mg once every 4 months.

Exclusion criteria:

Requests may not be approved for the following:

- Concomitant therapy with Evrysdi® (risdiplam);
- Product use for non-FDA approved indications as Spinraza is considered investigational/experimental when used for all other conditions, including but not limited to non-5q SMA or any other types of SMA not specified.
- Doses, durations, or dosing intervals that exceed FDA maximum limits for any FDA-approved indication or are not supported by industry-accepted practice guidelines or peer-reviewed literature for the relevant off-label use;
- Individuals with significant known risk factors unless the record provides an assessment of clinical benefit that outweighs the risk;

Initial authorization is provided for up to 6 months with a limitation of a maximum of 6 doses during the first year and 3 doses annually thereafter.

Reauthorizations will require medical chart documentation that the individual has been seen within the past 6 months and that markers of disease are improved by therapy:

- Individual does not require use of invasive ventilatory support or use of non-invasive ventilator support (BiPAP) for more than 16 hours per day as a result of advanced SMA disease; **AND**
- Documentation is provided that individual has clinically significant improvement in spinal muscular atrophy-associated signs and symptoms (i.e., progression, stabilization, or decreased decline in motor function) compared to the predicted natural history trajectory of disease or as supported by clinically significant improvement of motor function as measured by exams(HINE, HFMSE, ULM, CHOP-INTEND)

Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

U.S. Food and Drug Administration:
This section is to be used for informational purposes. FDA approval alone is not a basis for coverage.

SPINRAZA® is a survival motor neuron-2 (SMN2)-directed antisense oligonucleotide indicated for the treatment of spinal muscular atrophy (SMA) in pediatric and adult individuals.

References:

FDA approves first drug for spinal muscular atrophy | FDA. (2016, December 23). US Food and Drug Administration. Retrieved June 20, 2022, from https://www.fda.gov/news-events/press-announcements/fda-approves-first-drug-spinal-muscular-atrophy
Figueiredo, M. (2021, December 20). *Spinraza (Nusinersen) for the Treatment of People With SMA*. SMA News Today. Retrieved June 20, 2022, from https://smanewstoday.com/spinraza-nusinersen/
Nusinersen (Lexi-Drugs). (2022, June 20). Lexicomp. Retrieved June 20, 2022, from https://online.lexi.com/lco/action/doc/retrieve/docid/patch_f/6357166?cesid=31WNzAoqM83&searchUrl=%2Flco%2Faction%2Fsearch%3Fq%3Dspinraza%26t%3Dname%26acs%3Dfalse%26acq%3Dspinraza#doa
SPINRAZA (nusinersen). (2020, June). Spinraza. Retrieved June 20, 2022, from https://www.spinraza.com/content/dam/commercial/spinraza/caregiver/en_us/pdf/spinraza-prescribing-information.pdf
SPINRAZA (nusinersen). (2020, June). Accessdata.fda.gov. Retrieved June 20, 2022, from https://www.accessdata.fda.gov/drugsatfda_docs/label/2020/209531s010lbl.pdf
Vaidya, S., & Boes, S. (2018, December). Measuring quality of life in children with spinal muscular atrophy: a systematic literature review. *Quality of life research : an international journal of quality of life aspects of treatment, care and rehabilitation*, *27*(12), 3087-3094. PubMed.gov. https://doi.org/10.1007/s11136-018-1945-x

Criteria History/ Revision Information:

Date	**Summary of Changes**
June 2022	Criteria for use summary developed by Ascension Medical Specialty Prior Authorization Team
July 2022	Criteria for use summary approved by Ascension Neurology Expert Review Panel
July 2022	Criteria for use summary approved by Ascension Therapeutic Affinity Group

If you have questions, call 833-980-2352 to speak to a member of the Ascension Rx prior authorization team or email your questions to smarthealthspecialty@ascension.org.