Physician Administered Drug Prior Authorization Criteria for Spinraza®

Version, 1-21-20
Spinraza® Nusinersen

12mg/mL (2.4mg/mL) clear and colorless solution in a single-dose vial for intrathecal administration

Mechanism of Action:
"SPINRAZA® is an antisense oligonucleotide (ASO) designed to treat SMA caused by mutations in chromosome 5q that lead to SMN protein deficiency. Using in vitro assays and studies in transgenic animal models of SMA, SPINRAZA® was shown to increase exon 7 inclusion in SMN2 messenger ribonucleic acid (mRNA) transcripts and production of full-length SMN protein." [1]

Indication:

SPINRAZA® is a survival motor neuron-2 (SMN2)-directed antisense oligonucleotide indicated for the treatment of spinal muscular atrophy (SMA).[1]

Dosage Form/Strength:

- Injection: 12mg/5mL (2.4mg/mL) as a clear and colorless solution in a single-dose vial.

Criteria for Approval:

1. Initial Authorization Request must include:
    - Monitoring plan
    - Previous therapies trialed and the nature of the failure
    - Complete medication regimen
2. The patient will be treated by a specialist. **AND, ALL OF THE FOLLOWING**
    a. Diagnosis was confirmed by genetic testing
    b. Type I, II, or III SMA
    c. Patient is 15 years old or younger at initiation of treatment
    d. Patient has not received Zolgensma™

e. Prescriber agrees to do a platelet count and coagulation test before each dose. Patient must have a platelet count of > 50,000 cells per microliter
f. Prescriber agrees to do quantitative spot urine testing before each dose
g. Obtain an age/SMA Type appropriate baseline motor milestone score from **ONE** the following assessments:
    - HINE (appropriate for children 2 to 24 months)
    - CHOP-INTEND (appropriate for infants with SMA Type 1 and children with an infant's repertoire of motor skills)
    - Revised Upper Limb Module (RULM) (appropriate for non-ambulatory children over 30 months)
    - Hammersmith Functional Motor Scale Expanded (HFMSE) (appropriate for later-onset SMA in persons over 24 months)

Criteria for Reauthorization Approval:

1. Age/SMA Type appropriate test for follow-up has been performed and shows a response to therapy either by improvement or stabilization (no decline in mobility/motor function).

2. Provider has performed all required lab tests, and these have shown no contraindication to continuation of therapy.

Criteria for Denial:

1. Lack of response **OR,**
2. Missed doses **OR,**
3. Known hypersensitivity to Spinraza® or any of its excipients.

Length of Authorization:

1. Initial coverage may be approved for 7 months through the 4 loading doses and 1 maintenance dose.
2. Subsequent re-authorizations may be approved for an additional year or 3 doses.

Quantity Limit:

1. The dispensing limit is 1 vial per 120 days.
2. Quantity limit does not apply for loading doses which include 4 doses (4 vials), administering 1 vial every 14 days for 3 doses, then the 4th dose 30 days following the 3rd dose and a maintenance dose 4 months after 4th dose.

CAUTIONS:

- Spinraza® must be administered by a healthcare professional trained in lumbar puncture and intrathecal injection.
- Spinraza® must be prescribed at no more than 12mg per dose administration.
- Close monitoring and adequate follow-up is required in both circumstances for the safety of the patient.

- The member will be monitored at baseline and at each dose, with for the following laboratory tests: platelet count, coagulation testing, and quantitative spot urine protein testing. Member is monitored for bleeding complications.

References / Footnotes:

1. Spinraza® [package insert]. Cambridge, MA: Biogen Inc. 6/2019, Accessed January 19, 2020.
2. Bodamer, OA. Spinal muscular atrophy. Dashe,J, ed. UpToDate. Waltham, MA: UpToDate Inc. http://www.uptodate.com (Accessed on January 19, 2020)
3. Lexicomp Online®, Lexi-Drugs®, Hudson, Ohio: Lexi-Comp, Inc.; January 19, 2020.