Montana Healthcare Programs
Physician Administered Drug Coverage Criteria

SPINRAZA® (nusinersen)

I. Medication Description

Spinraza® is a survival motor neuron-2 (SMN-2)-directed antisense oligonucleotide indicated for:

- Treatment of spinal muscular atrophy (SMA) in pediatric and adult patients.

II. Position Statement

Coverage is determined through a prior authorization process **that must include** supporting clinical documentation for each request.

III. Initial Coverage Criteria

Member must meet all the following criteria:

- Member must have a diagnosis of Spinal Muscular Atrophy Type 1, 2, or 3 (SMA1, SMA2, or SMA3) confirmed by genetic testing.
- Genetic testing has confirmed chromosome 5q homozygous deletions or dysfunctional point mutations of the SMN1 gene and 2 to ≤4 copies of SMN2 gene.
- Member must not have permanent ventilator dependence.
- Spinraza® is prescribed by a neurologist.
- Prescriber must submit documentation of a baseline motor function milestone evaluation using at least one of the following age-appropriate screening tools:
    - HINE-2 (Hammersmith Infant Neurological Exam Part 2) - appropriate for children 2 to 24 months of age
    - CHOP-INTEND (Children's Hospital of Philadelphia Infant Test of Neuromuscular Diseases)- appropriate for infants, children, and older people with an infant's repertoire of motor skills
    - HFMSE (Hammersmith Functional Motor Scale Expanded)- appropriate for individuals over 24 months of age with later-onset SMA (Type 2 or Type 3)
    - RULM (Revised Upper Limb Module Test)- appropriate for assessing upper limb function of ambulatory and non-ambulatory individuals
    - 6MWT (6 Minute Walking Test)- appropriate for ambulatory patients with later-onset SMA (Type 2 or Type 3).
- Provider attests that the following laboratory tests will be performed at baseline and prior to each administration of Spinraza®: platelet count, coagulation test, quantitative spot urine protein test.
- Member has not previously received Zolgensma®, or member has previously received Zolgensma® and has experienced a worsening in clinical status.
- Member is not concurrently using Evrysdi™.

IV. Renewal Coverage Criteria

Member must meet all the following criteria:

- Member has been adherent to Spinraza®.
- Member has experienced a positive clinical response, as demonstrated by improvement or maintenance of motor skills as compared to pre-treatment baseline using at least one of the following age appropriate screening tools:
    - HINE-2 (Hammersmith Infant Neurological Exam Part 2) - appropriate for children 2 to 24 months of age

- CHOP-INTEND (Children's Hospital of Philadelphia Infant Test of Neuromuscular Diseases)- appropriate for infants, children, and older people with an infant's repertoire of motor skills
- HFMSE (Hammersmith Functional Motor Scale Expanded)- appropriate for individuals over 24 months of age with later-onset SMA (Type 2 or Type 3)
- RULM (Revised Upper Limb Module Test)- appropriate for assessing upper limb function of ambulatory and non-ambulatory individuals
- 6MWT (6 Minute Walking Test)- appropriate for ambulatory patients with later-onset SMA (Type 2 or Type 3).

- Provider attests that the following laboratory tests are being performed prior to each administration of Spinraza®: platelet count, coagulation test, and quantitative spot urine test.
- Member has not received Zolgensma®, or member has previously received Zolgensma® and has experienced a worsening in clinical status.
- Member is not concurrently using Evrysdi™.

V. Quantity Limitations

Max of 12mg/dose intrathecally, as follows:

- The first 3 loading doses should be administered at 14-day intervals. The 4th loading dose should be administered 30 days after the 3rd dose.
- A maintenance dose should be administered once every 4 months thereafter.

VI. Coverage Duration

Initial approval duration: 6 months (total of first 4 loading doses and 1 maintenance dose)
Renewal approval duration: 1 year