Nusinersen (Spinraza) – PA Criteria

HCPC: J2326

Spinraza is an antisense oligonucleotide indicated for the treatment of spinal muscular atrophy (SMA). It is covered by South Dakota Medicaid following prior authorization when the patient meets the following criteria:

** All requests under this policy require SD medical director review **in addition** to meeting specified criteria below **

- **Initial Therapy (must meet all):**
  - Therapy is prescribed by a neurologist with expertise in the treatment of SMA
  - Individual has an SMA diagnosis with submitted documentation (e.g., chart notes, laboratory values) confirming the mutation or deletion of genes in chromosome 5q resulting in **one** of the following:
    - Homozygous gene deletion or mutation of SMN1 gene (e.g., homozygous deletion of exon 7 at locus 5q13)
    - Compound heterozygous mutation of SMN1 gene (e.g., deletion of SMN1 exon 7 [allele 1] and mutation of SMN1 [allele 2])
  - Individual has ≤3 copies of SMN2 gene
  - Individual is not dependent on **either** of the following:
    - Invasive ventilation or tracheostomy
    - Use of non-invasive ventilation beyond use for naps and nighttime sleep
  - Baseline functional status is documented by **one** of the following:
    - Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorder score (CHOP INTEND)
    - Hammersmith Infant Neurological Exam (HINE)
    - Hammersmith Functional Motor Scale Expanded (HFMSE)
  - Therapy is not prescribed concurrently with Evrysdi or other SMN modifying therapies
  - Individual has not previously received gene replacement therapy for the treatment of SMA – request for coverage post gene replacement therapy will be determined on a case by case basis
  - Approval duration: 1 year (up to 4 doses)
- **Continuation of Therapy (must meet all):**
  - Individual continues to meet initial criteria
  - Submission of medical records (e.g., chart notes, laboratory values) with the most recent results (< 1 month prior to request) documenting a positive clinical response from pretreatment baseline status to Spinraza therapy as demonstrated by the initial test used to establish baseline motor ability unless that test is no longer appropriate for the patient
  - Improvement standards must meet **one** of the following:
    - Maintenance or improvement in score from pretreatment baseline
    - Patient has achieved and maintained any new motor milestone from pretreatment baseline when they would otherwise be unexpected to do so
  - Approval duration: 1 year

Spinraza is not covered for:

- Spinal muscular atrophy without chromosome 5q mutations or deletions
- Routine concomitant treatment of SMA in patients who have recently received gene replacement therapy with unknown clinical response