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

### Coverage Guidelines

1. **Patient Population**
   - AllWays Health Partners may authorize coverage of Spinraza (nusinersen) for members, when ALL of the following criteria are met:
     - Members have a documented diagnosis of spinal muscular atrophy (SMA) type 1, 2, or 3 confirmed by molecular genetic testing
     - Clinical documentation of baseline (pre-treatment) motor function skills has been submitted
     - Members have already established care with a SMA multidisciplinary care center
     - Members have none of the following: hospitalization for surgery or pulmonary event within past 2 months, active infection, brain or spinal cord disease, meningitis, implanted CSF shunt, treatment with another investigational drug <1 month of evaluation

2. **Prescribing**
   - Prescribed by neurologist with expertise in the management of SMA

3. **Dosing and Administration**
   - 4 loading doses: First 3 loading doses at 14 day intervals, 4th loading dose 30 days after 3rd dose
   - Maintenance dose every 4 months after the 4th loading dose
• Dose: 12 mg (5 mL) given intrathecally as bolus injection over 1-3 minutes using a spinal anesthesia needle
• Prior to administration, remove 5 mL of cerebral spinal fluid (CSF)
• Administered by attending neurologist experienced in administering intrathecal injections

4. Monitoring
• At baseline and prior to each dose, obtain a platelet count, coagulation test (i.e., prothrombin time, activated partial thromboplastin time) and quantitative spot urine protein test
• At each visit, assessment for improvement in clinical outcomes via motor function using HINE, CHOP-INTEND, HFMSE or other age-appropriate motor function scales

5. Duration of Therapy
• May be continued until disease progression or unacceptable toxicity (may require several months to a year for improvement in motor function to be seen)
• Discontinuation of drug to be determined based on age-appropriate performance on motor function and patient reported outcome scales using standardized instrument(s)

**Continuation of Therapy**
Reauthorization requires physician documentation of assessment of improvement in clinical outcomes via motor function using HINE, CHOP-INTEND, HFMSE or other age-appropriate motor function scale.

**Limitations**
1. Approvals will be granted for 12 months.

**References**

Review History
02/2017 – Reviewed by Clinical Experts
08/2017 – Revised (P&T approval)
11/2018 – Reviewed
03/18/2020 – Reviewed P&T Mtg

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