Spinraza (Nusinersen)

Spinraza will be covered when all of the criteria below are met, confirmed with supporting medical documentation.

I. Criteria for Initial Approval

- Diagnosis of SMA Type I, II, or III; AND
- Diagnosis by a neurologist with expertise in the diagnosis of SMA; AND
- Genetic testing confirming both:
    - 5q SMA homozygous gene deletion, homozygous gene mutation, or compound heterozygous mutation;
    - At least 2 copies of SMN2; AND
- Patient is not dependent on invasive ventilation or tracheostomy; AND
- Patient is not dependent on non-invasive ventilation beyond use for naps and nighttime sleep; AND
- Patients with Type II and III SMA must have some functional upper extremity use; AND
- Prescribed by a neurologist experienced in treating SMA; AND
- Baseline motor examination completed utilizing at least one of the following exams (based on patient age and motor ability) to establish baseline motor ability:
    - Hammersmith Infant Neurological Exam (HINE); or
    - Hammersmith Functional Motor Scale Expanded (HFMSE); or
    - Upper Limb Module Test (non-ambulatory); or
    - Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND)

II. Criteria for Continuation of Therapy

- All of the criteria for initial therapy in Section I. must be met; AND
- Repeat motor testing completed since the most recent Spinraza dose (and not more than 1 month prior to the next scheduled dose) using the same motor test done to establish baseline motor ability, unless it is determined that the original test is no longer appropriate; AND
- Repeat motor testing must document a response to treatment as defined by the following:
    - HINE:
        - Improvement or maintenance of previous improvement of at least 2 points (or max score of 4) in ability to kick (improvement in at least 2 milestones); OR
        - Improvement or maintenance of previous improvement of at least 1 point increase in motor milestones of head control, rolling, sitting,

crawling, standing or walking (consistent with improvement by at least 1 milestone); AND
        - Improvement or maintenance of previous improvement in more HINE motor milestones than worsening;

    - HFMSE:
        - Improvement or maintenance of improvement of at least a 3 point increase in score;
    - ULM:
        - Improvement or maintenance of previous improvement of at least 2 point increase in score;

    - CHOP-INTEND:
        - Improvement or maintenance of previous improvement of at least 4 point increase in score.

III. Dosing/Administration

Spinraza must be administered according to the current FDA labeling guidelines for dosage and timing. Spinraza must be administered intrathecally by a physician or other healthcare professional experienced in performing lumbar punctures.

- The recommended dosage is 12 mg (5 mL) per administration
- Initiate Spinraza treatment with 4 loading doses; the first three 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

IV. Length of Authorization

Spinraza will initially be preauthorized for 4 loading doses when criteria are met. Preauthorization is valid for 90 days. Each Spinraza maintenance dose (continuing therapy) must be preauthorized.

Maryland Medicaid considers Spinraza investigational and not medically necessary when the criteria above are not met and for all other indications.