Division: Pharmacy Policy	Subject: State of Florida's Agency for Health Care Administration's Prior Authorization Criteria
Original Development Date: Original Effective Date: Revision Date:	June 20, 2017 July 21, 2017, August 28, 2017, January 18, 2019, June 24, 2020, September 23, 2020, December 15, 2020, April 23, 2021

SPINRAZA® (nusinersen)

LENGTH OF AUTHORIZATION: 5 doses/8 months

REVIEW CRITERIA:

- Confirmed diagnosis of spinal muscular atrophy (SMA) confirmed by genetic testing.
  1. Documentation of genetic testing confirming either two or three copies of SMN2 gene;

     OR

     Documentation of genetic testing confirming four copies of SMN2 gene with symptomatology of SMA.
  2. Genetic testing confirms the presence of one of the following (a, b or c):
     a. Homozygous deletions of SMN1 gene (e.g., absence of the SMN1 gene);
     b. Homozygous mutation in the SMN1 gene (e.g., biallelic mutations of exon 7);
     c. Compound heterozygous mutation in the SMN1 gene (e.g., deletion of SMN1 exon 7 (allele 1) and mutation of SMN1 (allele 2))
- Medication is prescribed or in consultation with a pediatric neuromuscular specialist or a neurologist specializing in SMA.
- Obtain baseline assessment motor milestone score from ONE of the following assessments:
  - Hammersmith Functional Motor Scale Expanded (HFMSE)
  - Hammersmith Infant Neurologic Exam (HINE)
  - Upper limb module (ULM) score
  - Revised upper limb module (RULM) score
  - Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND)
  - Six-minute walk test
- Platelet count, coagulation laboratory testing and quantitative spot urine protein testing are required at baseline and prior to each administration.
- Patient is not dependent on either of the following:
  - Invasive ventilation (for not more than 16 hours per day) or tracheostomy OR
  - Non-invasive ventilation for at least 12 hours per day
- Specifically, for older patients with SMA and scoliosis, the drug may only be authorized if the patient has:

  a. Scoliosis without spine surgery, or

b. Is post spine surgery with preserved window of accessibility, by intrathecal injection under fluoroscopic or ultrasound guidance if needed, or
c. Is post spine surgery (e.g., fusion) without window of accessibility with surgical placement of an indwelling catheter or establishment a new window for IT accessibility.

- Patient is not concurrently treated with Evrysdi.

CONTINUATION OF THERAPY:

LENGTH OF AUTHORIZATION: 8 months

- Patient met initial review criteria.
- Submission of most recent platelet count, coagulation laboratory testing and quantitative spot urine protein versus pretreatment baseline status.
- Patient must not have advanced SMA and is not dependent on either of the following:
  - Invasive ventilation (for not more than 16 hours per day) or tracheostomy OR
  - Non-invasive ventilation for at least 12 hours per day
- Documentation of a positive response to therapy and that the patient is responding to the medication as demonstrated by clinically significant improvement or maintenance of function from pretreatment baseline status (progression, stabilization, or decreased decline in motor function) from ONE of the following age appropriate assessments:
  - Hammersmith Functional Motor Scale Expanded (HFMSE)
  - Hammersmith Infant Neurologic Exam (HINE)
  - Upper limb module (ULM) score
  - Revised upper limb module (RULM) score
  - Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND)
  - Six-minute walk test
- Patient is not concurrently treated with Evrysdi.

DOSING:

- Given intrathecally, the recommended dose is 12 mg (5 mL) per administration.