Market Applicability/Effective Date														
Market	**FL & FHK**	**FL MMA**	**FL LTC**	**GA**	**KS**	**KY**	**LA**	**MD**	**NJ**	**NV**	**NY**	**TN**	**TX**	**WA**
Applicable	NA	N/A	N/A	N/A	N/A	N/A	N/A	X	N/A	N/A	N/A	N/A	N/A	N/A

*FHK- Florida Healthy Kids

Spinraza (Nusinersin)

APPROVAL CRITERIA

Requests for Spinraza (Nusinersin) may be approved when the following criteria are met:

I. Diagnosis of spinal muscular atrophy (SMA) Type 1 with symptom onset at < 6 months of age; **AND**
II. Diagnosis by a neurologist with expertise in the diagnosis of SMA; **AND**
III. Genetic testing confirming both:
   A. 5q SMA homozygous gene deletion, homozygous gene mutation, or compound heterozygous mutation; **AND**
   B. At least 2 copies of SMN2

AND

IV. Patient is not dependent on invasive ventilation or tracheostomy; **AND**
V. Patient is not dependent on non-invasive ventilation for greater than 12 hours in a 24 hour period;

AND

VI. Spinraza must be prescribed by a neurologist experienced in treating SMA; **AND**
VII. Spinraza must be given according to the current FDA labelling guidelines for dosage and timing; **AND**
VIII. Spinraza must be administered intrathecally by a physician or other healthcare professional experienced in performing lumbar punctures; **AND**

For Initial Therapy:

A. Medical records must be submitted documenting all of the above criteria; **AND**
B. Medical records must be submitted documenting a baseline motor examination utilizing at least one of the following exams (based on patient age and motor ability) to establish baseline motor ability:
   1. Hammersmith Infant Neurological Exam (HINE); **OR**
   2. Hammersmith Functional Motor Scale Expanded (HFMSE); **OR**
   3. Upper Limb Module Test (non-ambulatory); **OR**
   4. Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND); **AND**
C. Spinraza will be initially preauthorized for 4 loading doses when criteria are met.

For Continuing Therapy:

A. Each Spinraza maintenance dose must be preauthorized; **AND**
B. All of the criteria for initial therapy must be met; **AND**
C. Medical records must be submitted that document repeat motor testing since the most recent Spinraza dose (and not more than 1 month prior to the next

This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.

Market Applicability/Effective Date														
Market	**FL & FHK**	**FL MMA**	**FL LTC**	**GA**	**KS**	**KY**	**LA**	**MD**	**NJ**	**NV**	**NY**	**TN**	**TX**	**WA**
Applicable	NA	N/A	N/A	N/A	N/A	N/A	N/A	X	N/A	N/A	N/A	N/A	N/A	N/A

*FHK- Florida Healthy Kids

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**

D. Repeat motor testing must document a response to treatment as defined by one of the following:
    1. HINE
        a. 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**
        b. 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**
        c. Improvement or maintenance of previous improvement in more HINE motor milestones than worsening;

    OR
    2. HFMSE
        a. Improvement or maintenance of improvement of at least a 3 point increase in score; **OR**
    3. ULM
        a. Improvement or maintenance of previous improvement of at least 2 point increase in score; **OR**
    4. CHOP-INTEND
        a. Improvement or maintenance of previous improvement of at least 4 point increase in score.

Requests for Spinraza (Nusinersin) may not be approved when the criteria above are not met and for all other indications.

This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.