Missouri Care

'Ohana Health Plan, a plan offered by WellCare Health Insurance of Arizona

Staywell of Florida

Children's Medical Services Health Plan (CMS Health Plan)

Heritage Health

WellCare (Alabama, Arizona, Arkansas, California, Connecticut, Florida, Georgia, Illinois, Indiana, Louisiana, Maine, Michigan, Mississippi, Missouri, New Hampshire, New Jersey, New York, North Carolina, Ohio, South Carolina, Tennessee, Texas, Washington)

WellCare Heritage Health

WellCare Prescription Insurance

WellCare TexanPlus (Medicare – Dallas & Houston markets)

Spinraza®

Policy Number: HS-243

Original Effective Date: 4/6/2017

Revised Date(s): 6/1/2017; 6/23/2017; 1/4/2018; 7/12/2018; 6/6/2019; 9/5/2019; 2/17/2020

APPLICATION STATEMENT

The application of the Clinical Coverage Guideline is subject to the benefit determinations set forth by the Centers for Medicare and Medicaid Services (CMS) National and Local Coverage Determinations and state-specific Medicaid mandates, if any.

DISCLAIMER

The Clinical Coverage Guideline (CCG) is intended to supplement certain standard WellCare benefit plans and aid in administering benefits. Federal and state law, contract language, etc. take precedence over the CCG (e.g., Centers for Medicare and Medicaid Services [CMS] National Coverage Determinations [NCDs], Local Coverage Determinations [LCDs] or other published documents). The terms of a member's particular Benefit Plan, Evidence of Coverage, Certificate of Coverage, etc., may differ significantly from this Coverage Position. For example, a member's benefit plan may contain specific exclusions related to the topic addressed in this CCG. Additionally, CCGs relate exclusively to the administration of health benefit plans and are NOT recommendations for treatment, nor should they be used as treatment guidelines. Providers are responsible for the treatment and recommendations provided to the member. The application of the CCG is subject to the benefit determinations set forth by the Centers for Medicare and Medicaid Services (CMS) National and Local Coverage Determinations, and any state-specific Medicaid mandates. Links are current at time of approval by the Medical Policy Committee (MPC) and are subject to change. Lines of business are also subject to change without notice and are noted on www.wellcare.com. Guidelines are also available on the site by selecting the Provider tab, then "Tools" and "Clinical Guidelines".

BACKGROUND

Spinal Muscular Atrophy (SMA) disorders are characterized by degeneration of the anterior horn cells in the spinal cord and motor nuclei in the lower brainstem. These diseases are classified as types 1 through 4 depending upon the age of onset and clinical course. The incidence of spinal muscular atrophy ranges from 4 to 10 per 100,000 live births, and the carrier frequency of disease-causing SMN1 mutations ranges from 1/90 to 1/50.[1]

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- **SMA type 1 (infantile spinal muscular atrophy or Werdnig-Hoffmann disease)** is the most common and severe type of SMA. It typically presents in the neonatal period. Symptoms progress rapidly, and the majority of infants die before one year of age from respiratory failure.
- **SMA type 2 (intermediate form) and type 3 (Kugelberg-Welander disease)** have a less severe course. SMA 2 presents between three and 15 months of age. SMA 3, the least severe, typically presents at or after one year of age and progresses to a chronic course.
- **Adult onset of SMA (type 4)** usually presents in the second or third decade of life and is otherwise similar to SMA type 3.

Differential diagnosis of infantile SMA (types 0 and 1) includes rule out of other causes of floppy infants. Of particular importance are the following conditions:[1]

- Arthrogryposis multiplex congenita
- X-linked infantile spinal muscular atrophy
- Spinal muscular atrophy with respiratory distress type 1
- Congenital myasthenic syndromes
- Congenital myopathies
- Hypoxic-ischemic myelopathy
- Lysosomal acid maltase deficiency
- Prader-Willi syndrome
- Traumatic myelopathy
- Zellweger syndrome

Treatment for SMA was previously supportive and directed at providing nutrition and respiratory assistance as needed, and treating or preventing complications of weakness. Disease-modifying therapy is now available with the approval of nusinersen in the United States. Nusinersen is recommended for most infants with SMA. Published data on nusinersen in older children, adults, and patients with advanced disease is limited. Due to the uncertainty regarding potential long-term adverse effects of this therapy, individualized treatment decision in these patients is advised.[1]

Nusinersen

Spinraza® (nusinersen) was the first drug approved to treat pediatric and adult patients with SMA. The drug is an antisense oligonucleotide that is designed to increase the expression of the survival motor neuron protein which is deficient in SMA. Initial clinical trials showed that intrathecal nusinersen therapy was safe and tolerable, and there was a suggestion of clinical benefit. The FDA announced approval of Spinraza® December 2016.[1]

The most common adverse events associated with intrathecal Spinraza® treatment were respiratory tract infections and constipation. The prescribing label notes an increased risk for thrombocytopenia, coagulation abnormalities, and renal toxicity. Thus, laboratory testing for platelet count, prothrombin time, activated partial thromboplastin time, and quantitative spot urine protein is recommended at baseline and prior to each dose.[1]

Administration. Spinraza® is administered by intrathecal (spinal) injection; each dose is 12 mg per 5 mL supplied in a single vial. Treatment is initiated with four loading doses; the first three loading doses are given at 14 day intervals, while the fourth loading dose is given 30 days after the third. There the initial treatment, a maintenance dose is given once every 4 months. The cost of each dose is listed as $125,000.[1]

Prior to administration, the medication must warm to room temperature. For the procedure the patient will have 5 mL of cerebrospinal fluid removed and the medication is given by intrathecal bolus injection over 1 to 3 minutes.[2]

After Spinraza® is administered in an outpatient setting, some patients may require additional monitoring and/or management in a hospital facility. For certain patients, providers may also consider administering Spinraza® in a hospital inpatient setting.[4]

Side Effects. Members should be instructed to call their provider if they have any of the following:[3]

- Signs of an allergic reaction, like rash; hives; itching; red, swollen, blistered, or peeling skin with or without fever; wheezing; tightness in the chest or throat; trouble breathing or talking; unusual hoarseness; or swelling of the mouth, face, lips, tongue, or throat

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- Signs of bleeding like throwing up blood or throw up that looks like coffee grounds; coughing up blood; blood in the urine; black, red, or tarry stools; bleeding from the gums; vaginal bleeding that is not normal; bruises without a reason or that get bigger; or any bleeding that is very bad or that you cannot stop.
- Signs of kidney problems like unable to pass urine, change in how much urine is passed, blood in the urine, or a big weight gain
- Fever or chills
- Change in color of sputum
- Cough
- Ear pain
- Very bad headache
- Signs of a common cold
- Hard stools (constipation)
- Headache
- Back pain

The most common adverse reactions that occurred in the controlled study were lower respiratory infection and constipation. Because patients in the controlled study were infants, adverse reactions that are verbally reported could not be assessed in this study. In the open-label studies, the most common adverse events in later onset patients were headache, back pain and post lumbar puncture syndrome.[4]

Thrombocytopenia and Coagulation Abnormalities and Renal Toxicity did occur in a small number of trial participants. Baseline and follow up labs are required prior to and following each dose administration of Spinraza®.[4]

Platelet count should not fall below 50,000 cells per microliter. Urinary protein concentration should not be greater than 0.2g/L.[8]

Safety in Specific Populations

Pregnancy. There is no adequate data on the developmental risk associated with the use of SPINRAZA in pregnant women. The background risk of major birth defects and miscarriage for the indicated population is unknown. [4]

Lactation. There is no data on the presence of nusinersen in human milk, the effects on the breastfed infant, or the effects of the drug on milk production.[4]

Pediatric Use. The safety and effectiveness of Spinraza in pediatric patients from newborn to 17 years has been established through animal studies and clinical trials.[4]

Geriatric Use. SMA is largely a disease of children and young adults; therefore, there is no geriatric experience with Spinraza.[4]

Care Plan Checklist[4]

Determine if any ancillary services may be needed to support administration via intrathecal injection:

- Anesthesia
- Lumbar puncture (separate procedure)
- Ultrasound
- Fluoroscopy
- Other

Evaluate the need for and the feasibility of an outpatient observation stay post injection

- Observation stay as a possibility in lieu of inpatient admission

Select the setting and the site of care for Spinraza® administration

- Outpatient Setting
- Inpatient Setting
- Other facility
- Hospital outpatient on-campus facility

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- Other Setting
- Hospital outpatient off-campus clinic
- Inpatient hospital facility
- Hospital-based ASC
- Freestanding ASC
- Physician office

Identify which providers/provider practice groups will offer professional services related to Spinraza® administration

- Neurology
- Radiology
- Anesthesiology
- Other

Document the administration plan for the dosing schedule

- Dates of loading doses
- Dates of maintenance doses (if applicable)

Coordinate with the patient and his or her caregiver to confirm the care plan and to help set appropriate expectations

- Treatment process and related timelines
- Current payer coverage situation and any anticipated changes
- Potential financial assistance needs
- SMA360° support services available for patients and their families

Suggest SMA360° support services, which may be available to help the patient's family understand and navigate the treatment process

- Provide the member's family with the SPINRAZA Start Form; assist them in completing the patient portion, and review caregiver consent.
- Your practice or facility should complete the HCP portion of the SPINRAZA Start Form. Be sure to include the provider's signature in the Prescriber Authorization section. Fax the completed Start Form to 1-888-538-9781
- If signed consent is provided, advise the patient's family that a FAM from Biogen will assist them in coordinating the logistics of treatment, such as insurance and financial considerations, if needed

For patients who may require additional monitoring after Spinraza® administration, additional outpatient observation stay for up to 48 hours may be approved.

Some patients who receive Spinraza® may face restrictions from their commercial and/or Medicaid payers because the provider and/or the service facility is out of network or out of state. It is important to recognize that, for those instances, waivers or exceptions can be granted on the grounds of medical necessity.

- Verify the state and/or network participation status for the physician(s) and/or facility involved in the administration of Spinraza®
- Investigate and record the patient OOP cost implications for out-of-state and/or out-of-network providers
- Find out if there is an exception process for patients seeking care out of state and/or out of network

There may be cases where your patient has multiple payers that provide benefit coverage, such as a commercial health plan and Medicaid.

- In the case of multiple payers, it is important to establish during the Benefit Investigation which payer is first, which is secondary, and which is tertiary, if needed
- Once you have established the order of benefits, follow the instructions from each payer regarding coordination of benefits for reimbursement/payment methodology

Specific coding and billing requirements may vary by payer, particularly for claims with a miscellaneous J-code

- Find out if additional information is required for claims with a miscellaneous J-code and where to include it on the claim

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- Clarify the requirements for reporting an NDC number in a medical claim

Identify specific documentation that must be submitted with the request	Determine the preauthorization/prior authorization coverage parameters
• Letter of medical necessity • Chart notes • Specific payer preauthorization/prior auth form • Spinraza® Prescribing Information • Relevant literature (published standards of care) • Clinical documentation related to the disease, including: o Diagnostic evidence of SMA (eg genetic testing) o Clinical presentation, duration of symptoms o Current supportive care management o Care plan o Other relevant aspects of patient history	• Number of doses • Time limits of authorization • Diagnosis limitations • Submission requirements

POSITION STATEMENT

Applicable To:

- ☑ Medicaid – All Markets (Excluding KY)
- ☑ Children's Medical Services Health Plan (CHIP)
- ☑ Medicare – All Markets

Exclusions

Spinraza® **is considered experimental and investigational** for spinal muscular atrophy without chromosome 5q mutations or deletions.

Spinraza® **is not considered medically necessary and not a covered benefit** for members who have previously been treated with Zolgensma®.

Coverage

Spinraza® **is considered medically necessary** when ALL of the following criteria is met:

Initial Therapy
Note: Initial therapy pertains to members receiving the drug for the first time.

1. Member must have one of the following:
   a. Diagnosis of spinal muscular atrophy type I, II, or III by a neurologist trained in diagnosing SMA; **OR**
   b. Diagnosis of spinal muscular atrophy type I, II, or III by a physician in consultation with a neurologist trained in diagnosing SMA.

AND

2. Submission of medical records (e.g., chart notes, laboratory values) confirming both of the following:

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a. The mutation or deletion of genes in chromosome 5q resulting in one of the following:
   i. Homozygous gene deletion or mutation (e.g., homozygous deletion of exon 7 at locus 5q13); **OR**
   ii. Compound heterozygous mutation (e.g., deletion of SMN1 exon 7[allele 1] and mutation of SMN1 [allele 2])

AND

b. Patient has at least 2 copies of SMN2.

AND

~~3.~~ Patient is not dependent on either of the following:
   a. Invasive ventilation or tracheostomy
   b. Non-invasive ventilation (i.e. BiPAP) for more than 6 hours per day during wakeful hours.

AND

4. Submission of medical records (e.g., chart notes, laboratory results) of the baseline exam of at least one of the following exams (based on patient age and motor ability) to establish baseline motor ability:
   a. Hammersmith Infant Neurological Exam (HINE) (infant to early childhood); **OR**
   b. Hammersmith Functional Motor Scale Expanded (HFMSE); **OR**
   c. Upper Limb Module (ULM) Test (Non ambulatory); **OR**
   d. Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND).[6,8]

AND

5. Member's baseline labs were submitted and member has no evidence of thrombocytopenia or renal impairment.

AND

6. One of the following:
   a. Spinraza is being prescribed by a neurologist trained in diagnosing SMA; **OR**
   b. Spinraza is prescribed by a physician in consultation with a neurologist trained in diagnosing SMA.

AND

7. Spinraza® will be administered intrathecally by, or under the direction of, healthcare providers experienced in performing lumbar punctures.

AND

8. Spinraza® dosing for SMA is in accordance with the United States Food and Drug Administration approved labeling (maximum dosing of 12mg for each loading dose).

AND

9. Initial authorization will be for no more than 4 loading doses.

Continuation of Therapy

Note: Continued therapy is for any treatment following the initial administration regardless of whether or not the initial administration was approved by another health plan.

1. One of the following:
   a. Diagnosis of spinal muscular atrophy type I, II, or III by a neurologist trained in diagnosing SMA; **OR**
   b. Diagnosis of spinal muscular atrophy type I, II, or III by a physician in consultation with a neurologist trained in diagnosing SMA.

AND

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2. Submission of medical records (e.g., chart notes, laboratory values) confirming both of the following:
   a. The mutation or deletion of genes in chromosome 5q resulting in one of the following:
      i. Homozygous gene deletion or mutation (e.g., homozygous deletion of exon 7 at locus 5q13); **OR**
      ii. Compound heterozygous mutation (e.g., deletion of SMN1 exon 7[allele 1] and mutation of SMN1 [allele 2])

   AND

   b. Patient has at least 2 copies of SMN2.

AND

3. Patient is not dependent on either of the following: (Does not apply to Nebraska Medicaid)
   a. Invasive ventilation or tracheostomy
   b. Non-invasive ventilation (i.e. BiPAP) for more than 6 hours per day during wakeful hours.

AND

4. 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 at least one of the following exams:
   a. HINE milestones:
      i. One of the following:
         (i) Improvement or maintenance of previous improvement of at least 2 point (or maximal score) increase in ability to kick; **OR**
         (ii) Improvement or maintenance of previous improvement of at least 1 point increase in any other HINE milestone (e.g., head control, rolling, sitting, crawling, etc.), excluding voluntary grasp.

   AND

      ii. One of the following:
         (i) The patient exhibited improvement, or maintenance of previous improvement in more HINE motor milestones than worsening, from pretreatment baseline (net positive improvement); **OR**
         (ii) Achieved and maintained any new motor milestones when they would otherwise be unexpected to do so (e.g., sit unassisted, stand, walk).

   OR one of the following (b, c, or d):

   b. HFMSE: One of the following:
      i. Improvement or maintenance of previous improvement of at least a 3 point increase in score from pretreatment baseline; **OR**
      ii. Patient has achieved and maintained any new motor milestone from pretreatment baseline when they would otherwise be unexpected to do so.

   OR

   c. ULM: One of the following:
      i. Improvement or maintenance of previous improvement of at least a 2 point increase in score from pretreatment baseline; **OR**
      ii. Patient has achieved and maintained any new motor milestone from pretreatment baseline when they would otherwise be unexpected to do so.

   OR

   d. CHOP INTEND: One of the following:
      i. Improvement or maintenance of previous improvement of at least a 4 point increase in score from pretreatment baseline; **OR**
      ii. Patient has achieved and maintained any new motor milestone from pretreatment baseline when they would otherwise be unexpected to do so.

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AND

5. Member's baseline labs were submitted and member has no evidence of thrombocytopenia or renal impairment.

AND

6. One of the following:
   a. Spinraza® is prescribed by a neurologist trained in diagnosing SMA; **OR**
   b. Spinraza® is prescribed by a physician in consultation with a neurologist trained in diagnosing SMA.

AND

7. Spinraza® is to be administered intrathecally by, or under the direction of, healthcare professionals experienced in performing lumbar punctures.

AND

8. Spinraza® dosing for SMA is in accordance with the United States Food and Drug Administration approved labeling (maximum dosing of 12mg every 4 months, starting 4 months after the last loading dose).

AND

9. Reauthorization will be for no more than 3 maintenance doses (12 months).

Florida[13]

Spinraza® **is considered medically necessary** when ALL of the following criteria are met:

Initial Therapy
Note: Initial therapy pertains to members receiving the drug for the first time.

LENGTH OF AUTHORIZATION: 5 doses/8 months

1. Confirmed diagnosis of spinal muscular atrophy (SMA) confirmed by genetic testing.
   A. Documentation of genetic testing confirming either two or three copies** of SMN2 gene.
   B. Genetic testing confirms the presence of one of the following (a, b or c):
      i. Homozygous deletions of SMN1 gene (e.g., absence of the SMN1 gene);
      ii. Homozygous mutation in the SMN1 gene (e.g., biallelic mutations of exon 7);
      iii. Compound heterozygous mutation in the SMN1 gene (e.g., deletion of SMN1 exon 7
      iv. (allele 1) and mutation of SMN1 (allele 2))

2. Medication is prescribed or in consultation with a pediatric neuromuscular specialist or a neurologist specializing in SMA.

3. Obtain baseline assessment motor milestone score from ONE of the following assessments:
   A. Hammersmith Functional Motor Scale Expanded (HFMSE)
   B. Hammersmith Infant Neurologic Exam (HINE)
   C. Upper limb module (ULM) score
   D. Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP
   E. INTEND)
   F. Six-minute walk test

4. Platelet count, coagulation laboratory testing and quantitative spot urine protein testing are required at baseline and prior to each administration.

5. Patient is not dependent on either of the following:

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A. Invasive ventilation (for not more than 16 hours per day) or tracheostomy OR
B. Non-invasive ventilation for at least 12 hours per day

6. Specifically, for older clients with SMA and scoliosis, the drug may only be authorized if client 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.

Continuation of Therapy

LENGTH OF AUTHORIZATION: 8 months

1. Submission of most recent platelet count, coagulation laboratory testing and quantitative spot urine protein versus pretreatment baseline status.

2. Documentation of positive response to therapy such as:
   A. Documentation that the patient is responding to the medication as demonstrated by clinically significant improvement or maintenance of function from pretreatment baseline status using the same exam as performed at baseline assessment (progression, stabilization, or decreased decline in motor function):

      i. HFMSE: One of the following:
         - Patient has achieved and maintained any new motor milestone from pretreatment baseline when they would otherwise be unexpected to do so; **OR,**
         - Improvement or maintenance of previous improvement of at least a three-point increase in score from pretreatment baseline.

      OR

      ii. HINE milestones: One of the following
         - Improvement or maintenance of previous improvement of at least two-point (or maximal score) increase in ability to kick; **OR,**
         - Improvement or maintenance of previous improvement of at least one-point increase in any other HINE milestone (e.g., head control, rolling, sitting, crawling, etc.), excluding voluntary grasp.

      AND

      One of the following:
         - The patient exhibited improvement, or maintenance of previous improvement in more HINE motor milestones than worsening, from pretreatment baseline (net positive improvement); **OR,**
         - Achieved and maintained any new motor milestones when they would otherwise be unexpected to do so (e.g., sit unassisted, stand, walk).

      OR

      ULM: One of the following:
         - Improvement or maintenance of previous improvement of at least a two-point increase in score from pretreatment baseline; **OR,**
         - Patient has achieved and maintained any new motor milestone from pretreatment baseline when they would otherwise be unexpected to do so.

      OR

      CHOP INTEND: One of the following:
         - Improvement or maintenance of previous improvement of at least a four-point increase in score from pretreatment baseline; **OR,**
         - Patient has achieved and maintained any new motor milestone from pretreatment baseline when they would otherwise be unexpected to do so.

      OR

         - Six-minute walk test

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CODING

Considerations for Administration. Spinraza® is administered intrathecally by, or under the direction of, a healthcare provider with experience performing lumbar punctures. In addition, providers can consider the following services for the administration of SPINRAZA, as needed:[4]

- Sedation as indicated by the clinical condition of the patient
- Ultrasound or other imaging techniques to guide intrathecal administration of SPINRAZA, particularly in younger patients

Covered HCPCS Codes

J2326 Injection, nusinersen, 0.1 mg

Covered ICD-10-PCS Codes

3E0R3GC Introduction of Other Therapeutic Substance into Spinal Canal, Percutaneous Approach
BR13YZZ Fluoroscopy of Lumbar Disc(s) using Other Contrast
BR49ZZZ Ultrasonography of Lumbar Spine

Covered ICD-10-CM Codes

G12.0 Infantile spinal muscular atrophy, type I [Werdnig-Hoffman]
G12.1 Other inherited spinal muscular atrophy
G12.8 Other spinal muscular atrophies and related syndromes
G12.9 Spinal muscular atrophy, unspecified

Note that *ICD-10-CM* code G12.1 includes the following:

- Childhood form, type II spinal muscular atrophy
- Juvenile form, type III spinal muscular atrophy [Kugelberg-Welander]

Coding information is provided for informational purposes only. The inclusion or omission of a CPT, HCPCS, or ICD-10 code does not imply member coverage or provider reimbursement. Consult the member's benefits that are in place at time of service to determine coverage (or non-coverage) as well as applicable federal / state laws.

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MEDICAL POLICY COMMITTEE HISTORY AND REVISIONS

Date	Action
2/17/2020	• Approved by MPC. Added KY specific verbiage.
9/5/2019	• Approved by MPC. Added exclusion for member who have previously been treated with Zolgensma®.
6/6/2019	• Approved by MPC. No changes.
7/12/2018	• Approved by MPC. Semi-annual review; no changes.
1/4/2018	• Approved by MPC. Added FL specific criteria per ACHA guidelines.
6/23/2017	• Approved by MPC. Clarified language for changes approved at 6/1/2017 MPC meeting.
6/1/2017	• Approved by MPC. Amended criteria on mechanical ventilation dependency. Added NE specific age requirements.
4/6/2017	• Approved by MPC. New.

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