Louisiana Medicaid
Spinal Muscular Atrophy

The *Louisiana Uniform Prescription Drug Prior Authorization Form* should be utilized to request clinical authorization for spinal muscular atrophy agents (except Spinraza®).

The *Louisiana Medicaid Nusinersen (Spinraza®) Clinical Authorization Form* should be utilized to request clinical authorization for nusinersen (Spinraza®).

Additional Point-of-Sale edits may apply.

*These agents may have a **Black Box Warning** and may be subject to **Risk Evaluation and Mitigation Strategy (REMS)** under FDA safety regulations. Please refer to individual prescribing information for details.*

Nusinersen (Spinraza®)

Approval Criteria

- The recipient has a diagnosis of spinal muscular atrophy (SMA):
  - Type I, also known as infantile-onset or Werdnig-Hoffmann disease (ICD-10-CM G12.0), symptoms are present at birth or by 6 months of age, unable to sit without assistance; **OR**
  - Type II (ICD-10-CM G12.1), symptoms develop between 6 months and 12 months of age, able to sit unassisted but unable to stand or walk independently; **OR**
  - Type III, also known as Kugelberg-Welander disease (ICD-10-CM G12.1), usually diagnosed between early childhood and early adolescence, able to stand and walk independently but may lose this ability later in life; **AND**
- The diagnosis of SMA is confirmed with genetic testing; **AND**
- ~~The recipient is 16 years of age or younger at the initiation of treatment; **AND**~~
- The medication is prescribed by, or in consultation with, a physician who specializes in the treatment of spinal muscular atrophy; **AND**
- The recipient has not previously received treatment with Zolgensma (onasemnogene abeparvovec-xioi); **AND**
- **ONE** of the following motor milestone tests have been performed and the results are noted on the request form:
  - For recipients ≤ 2 years of age: Hammersmith Infant Neurological Examination Section 2 (HINE-2); **OR**
  - For ambulatory recipients ≥3 years of age: Hammersmith Functional Motor Scale Expanded (HFMSE); **OR**
  - For non-ambulatory recipients >3 years of age: Revised Upper Limb Module (RULM); **AND**
- If request is for a non-preferred agent – **ONE** of the following is required:
  - The recipient has had a *treatment failure* with at least one preferred product; **OR**
  - The recipient has had an *intolerable side effect* to at least one preferred product; **OR**

- The recipient has *documented contraindication(s)* to all of the preferred products that are appropriate to use for the condition being treated; **OR**
- There is *no preferred product that is appropriate* to use for the condition being treated; **AND**

- By submitting the authorization request, the prescriber attests to the following:
  - The prescribing information for the requested medication has been thoroughly reviewed, including any Black Box Warning, Risk Evaluation and Mitigation Strategy (REMS), contraindications, minimum age requirements, recommended dosing, and prior treatment requirements; **AND**
  - All laboratory testing and clinical monitoring recommended in the prescribing information have been completed as of the date of the request and will be repeated as recommended; **AND**
  - The recipient has no inappropriate concomitant drug therapies or disease states; **AND**
  - The recipient does not have a coexisting terminal condition or a condition with which the risk of nusinersen treatment outweighs the potential benefits.

Reauthorization Criteria

- Recipient continues to meet initial approval criteria; **AND**
- The prescriber states on the request that there has been a positive clinical benefit from nusinersen therapy as evidenced by:
  - improvement or maintenance of motor skills or ability to sit, crawl, stand or walk, or new motor milestone; **AND**
  - when considering all categories of motor milestones, the number of categories which show improvement is greater than the number that shows worsening.

Duration of initial authorization: 6 months
Duration of reauthorization: 12 months

Onasemnogene abeparvovec-xioi (Zolgensma®)

Approval Criteria

- The recipient has reached full-term gestational age (defined as 39 weeks 0 days) on the date of the request (documentation showing gestational age at birth [in weeks and days] must be provided with the request); **AND**
- The recipient is less than 2 years of age on the date of the request; **AND**
- This medication is prescribed by, or the request states that the medication is being prescribed in consultation with, a neurologist experienced in the treatment of SMA; **AND**
- The following are true and **stated on the request:**
  - The recipient has a diagnosis of spinal muscular atrophy (SMA) with bi-allelic mutations in the *survival motor neuron 1 (SMN1)* gene; **AND**
  - The recipient **DOES NOT HAVE advanced SMA** (e.g., complete paralysis of limbs, permanent ventilator dependence); **AND**

- The recipient **has never received a dose** of onasemnogene abeparvovec-xioi (Zolgensma®); **AND**
- The recipient has a baseline anti-AAV9 antibody titer ≤ 1:50, measured using an enzyme-linked immunosorbent assay (ELISA) **[date and results must be written on the request]**; **AND**

- If request is for a non-preferred agent **– ONE** of the following is required:
  - The recipient has had a *treatment failure* with at least one preferred product; **OR**
  - The recipient has had an *intolerable side effect* to at least one preferred product; **OR**
  - The recipient has *documented contraindication(s)* to all of the preferred products that are appropriate to use for the condition being treated; **OR**
  - There is *no preferred product that is appropriate* to use for the condition being treated; **AND**
- By submitting the authorization request, the prescriber attests to the following:
  - The prescribing information for the requested medication has been thoroughly reviewed, including any Black Box Warning, Risk Evaluation and Mitigation Strategy (REMS), contraindications, minimum age requirements, recommended dosing, prior treatment requirements (such as systemic corticosteroids) and required storage and handling procedures; **AND**
  - Where feasible, the recipient's vaccination schedule has been adjusted to accommodate concomitant corticosteroid administration prior to and following onasemnogene abeparvovec-xioi (Zolgensma®) infusion (seasonal RSV prophylaxis is not precluded); **AND**
  - All laboratory testing and clinical monitoring recommended in the prescribing information have been completed as of the date of the request and will be repeated as recommended; **AND**
  - The recipient has no concomitant drug therapies or disease states that limit the use of the requested medication and will not be receiving the requested medication in combination with any other medication that is contraindicated or not recommended per FDA labeling.

Duration of approval: 1 month

Risdiplam (Evrysdi®)

Approval Criteria

- The recipient has a diagnosis of spinal muscular atrophy (SMA):
  - Type I, also known as infantile-onset or Werdnig-Hoffmann disease (ICD-10-CM G12.0), symptoms are present at birth or by 6 months of age, unable to sit without assistance; **OR**
  - Type II (ICD-10-CM G12.1), symptoms develop between 6 months and 12 months of age, able to sit unassisted but unable to stand or walk independently; **OR**
  - Type III, also known as Kugelberg-Welander disease (ICD-10-CM G12.1), usually diagnosed between early childhood and early adolescence, able to stand and walk independently but may lose this ability later in life; **AND**

- The prescriber **states on the** request that the diagnosis of SMA has been confirmed with genetic testing; **AND**
- The medication is prescribed by, or **the request states** that it is in consultation with, a physician who specializes in the treatment of spinal muscular atrophy; **AND**
- The recipient has not previously received treatment with Zolgensma® (onasemnogene abeparvovec-xioi); **AND**
- The recipient is not using the requested medication concurrently with Spinraza® (nusinersen); **AND**
- The prescriber **states on the request** that at least one motor milestone test has been performed that is appropriate for the recipient's age and motor function; **AND**
- If request is for a non-preferred agent **– ONE** of the following is required:
    - The recipient has had a *treatment failure* with at least one preferred product; **OR**
    - The recipient has had an *intolerable side effect* to at least one preferred product; **OR**
    - The recipient has *documented contraindication(s)* to all of the preferred products that are appropriate to use for the condition being treated; **OR**
    - There is *no preferred product that is appropriate* to use for the condition being treated; **AND**
- By submitting the authorization request, the prescriber attests to the following:
    - The prescribing information for the requested medication has been thoroughly reviewed, including any Black Box Warning, Risk Evaluation and Mitigation Strategy (REMS), contraindications, minimum age requirements, recommended dosing, and prior treatment requirements; **AND**
    - All laboratory testing and clinical monitoring recommended in the prescribing information have been completed as of the date of the request and will be repeated as recommended; **AND**
    - The recipient has no concomitant drug therapies or disease states that limit the use of the requested medication and will not receive the requested medication in combination with any medication that is contraindicated or not recommended per FDA labeling; **AND**
    - The recipient does not have a coexisting terminal condition or a condition with which the risk of risdiplam treatment outweighs the potential benefits.

Reauthorization Criteria

- Recipient continues to meet initial approval criteria; **AND**
- The prescriber **states on the request** that the recipient is established on the medication with evidence of a positive response to therapy demonstrated by improvement or maintenance of motor milestone test that is appropriate for the recipient's age and motor function.

Duration of initial and reauthorization approval: 12 months

References

Evrysdi (risdiplam) [package insert]. South San Francisco, CA: Genentech, Inc. A Member of the Roche Group; October 2023. https://www.gene.com/download/pdf/evrysdi_prescribing.pdf

Spinraza (nusinersen) [package insert]. Cambridge, MA: Biogen; February 2023. https://www.spinraza-hcp.com/content/dam/commercial/specialty/spinraza/hcp/en_us/pdf/spinraza-prescribing-information.pdf

U.S. National Library of Medicine. Genetics Home Reference. (2018, September 25). Spinal Muscular Atrophy. https://ghr.nlm.nih.gov/condition/spinal-muscular-atrophy

Zolgensma (onasemnogene abeparvovec-xioi) [package insert]. Bannockburn, IL: AveXis, Inc.; October 2023. https://www.novartis.us/sites/www.novartis.us/files/zolgensma.pdf

Revision / Date	**Implementation Date**
Policy Created (Zolgensma®)	November 2019
Added requirement for motor milestone testing and modified reauthorization criteria to remove point-related increases in motor milestones / September 2019 (Spinraza®)	March 2020
Modified age to reflect updated prescribing information / September 2019 (Spinraza®)	March 2020
Included onasemnogene abeparvovec-xioi criteria statement / July 2020 (Spinraza®)	January 2021
Policy Created / August 2020 (Evrysdi®)	January 2021
Removed POS wording, formatting changes, updated references / May 2021 (Evrysdi®)	January 2022
Formatting changes, updated references / February 2022 (Zolgensma®)	July 2022
Removed age requirement, updated references / June 2022 (Evrysdi®)	October 2022
Modified motor milestone test requirements / August 2022 (Evrysdi®)	January 2023
Combined criteria for all spinal muscular atrophy agents / October 2023	January 2024
Removed age requirement at initiation of treatment for Spinraza® / November 2023	April 2024