TRUECARE

PHARMACY POLICY STATEMENT
Mississippi Medicaid

DRUG NAME	Spinraza (nusinersen)
BENEFIT TYPE	Medical
STATUS	Prior Authorization Required

Spinraza is a survival of motor neuron 2 (SMN2) splicing modifier initially approved by the FDA in 2020. It is indicated for the treatment of pediatric and adult patients with spinal muscular atrophy (SMA) with bi-allelic mutations in the survival motor neuron (SMN1) gene. Spinal muscular atrophy (SMA) is a genetic, autosomal recessive neuromuscular disorder caused by a defect in the survival of the motor neuron 1 (*SMN1*) gene. SMA is the leading genetic cause of infant mortality and affects approximately 1 in every 10,000 infants. There are multiple types of SMA, and the age of onset and severity of the disease varies with each type.

Spinraza (nusinersen) will be considered for coverage when the following criteria are met:

Spinal Muscular Atrophy (SMA)

For **initial** authorization:

1. Member's gestational age is 37 to 42 weeks for singleton births or 34 to 42 weeks for twins;
2. Medication must be prescribed by or in consultation with a neurologist; AND
3. Member has a diagnosis of SMA confirmed by genetic/newborn testing showing any of the following:
   a) Homozygous gene deletion of the survival motor neuron 1 (SMN1) gene (e.g., absence of SMN1 gene)
   b) Homozygous mutation of the SMN1 gene (e.g., biallelic mutation 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]); AND
4. Member has 2 to 4 copies of SMN2; AND
5. Member's documented oxygen saturation is ≥ 92% (awake or asleep) without any supplemental oxygen or respiratory support; AND
6. Member has documented ALL of the following:
   a) Prothrombin time, Activated partial prothrombin time;
   b) Platelet counts;
   c) Quantitative spot protein urine testing; AND
7. Member does not have shunt or central nervous system (CNS) catheter;
8. Member has no history of bacterial meningitis or viral encephalitis;
9. Member does not have prior treatment with Zolgensma (discontinuation of Spinraza prior to Zolgensma therapy is required and Spinraza will not be reauthorized after Zolgensma infusion).
10. Member will not be using concomitantly with Evrysdi.
11. **Dosage allowed/Quantity limit:** Initiate Spinraza treatment with 4 loading doses (12 mg (5 mL) per administration). 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. Quantity limit: 1 vial per 16 weeks

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:

1. Member has documentation of positive clinical improvement from pretreatment baseline status in spinal muscular atrophy-associated symptoms or maintenance (not worsening) of the disease state (e.g., decreased decline in motor function, increased ability to kick, increased in the motor milestones of head control, rolling, sitting, crawling, standing, or walking, etc.).

If all the above requirements are met, the medication will be approved for an additional 12 months.

TrueCare considers Spinraza (nusinersen) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
05/05/2017	New policy for Spinraza created.
06/11/2019	Concomitant used of Spinraza with Zolgensma will not be authorized. Spinraza must be discontinued before Zolgensma infusion. Spinraza will not be reauthorized after Zolgensma infusion.
07/11/2022	Transferred to new format. Updated references. Removed SMA typing. Removed baseline motor ability assessment scores.

References:

1. Spinraza [package insert]. Cambridge, MA; Biogen Inc.; June 2020.
2. Markowitz JA, Singh P, Darras BT. Spinal Muscular Atrophy: A Clinical and Research Update. Pediatric Neurology 46 (2012) 1-12.
3. Darras BT et al. An Integrated Safety Analysis of Infants and Children with Symptomatic Spinal Muscular Atrophy (SMA) Treated with Nusinersen in Seven Clinical Trials. CNS Drugs. 2019 Sep;33(9):919-932.
4. Dabbous O et al. Survival, Motor Function, and Motor Milestones: Comparison of AVXS-101 Relative to Nusinersen for the Treatment of Infants with Spinal Muscular Atrophy Type 1. Adv Ther. 2019 May;36(5):1164-1176.
5. Finkel RS et al; ENDEAR Study Group. Nusinersen versus Sham Control in Infantile-Onset Spinal Muscular Atrophy. N Engl J Med. 2017 Nov 2;377(18):1723-1732.
6. Finkel RS et al. Treatment of infantile-onset spinal muscular atrophy with nusinersen: a phase 2, open-label, dose-escalation study. Lancet. 2016 Dec 17;388(10063):3017-3026.
7. Farrar MA, et al. Emerging therapies and challenges in spinal muscular atrophy. Ann Neurol 2017;81(3):355–368.
8. De Sanctis R, et al. Developmental milestones in type I spinal muscular atrophy. Neuromusc Disord 2016;26(11):754–759.
9. Waldrop MA, et al. Current Treatment Options in Neurology—SMA Therapeutics. Curr Treatment Options Beurology. 2019;21(6):25.
10. Glascock, Jacqueline et al. Revised Recommendations for the Treatment of Infants Diagnosed with Spinal Muscular Atrophy via Newborn Screening Who have 4 Copies of SMN2. Journal of Neuro Dis. (2020) 97–100.

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