



AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name: Spinraza

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Effective Date: 11/27/2023

Last Review Date: 9/22/2023

Applies to:	<input checked="" type="checkbox"/> Illinois	<input type="checkbox"/> Florida	<input checked="" type="checkbox"/> Florida Kids
	<input type="checkbox"/> New Jersey	<input checked="" type="checkbox"/> Maryland	<input type="checkbox"/> Michigan
	<input checked="" type="checkbox"/> Pennsylvania Kids	<input checked="" type="checkbox"/> Virginia	<input checked="" type="checkbox"/> Kentucky PRMD

Intent:

The intent of this policy/guideline is to provide information to the prescribing practitioner outlining the coverage criteria for Spinraza under the patient's prescription drug benefit.

Description:

Spinraza is indicated for the treatment of spinal muscular atrophy (SMA) in pediatric and adult patients.

All other indications are considered experimental/investigational and not medically necessary.

Applicable Drug List:

Spinraza

Policy/Guideline:

Criteria for Initial Approval:

Spinal Muscular Atrophy (SMA)

A. Submission of the following information is necessary to initiate the prior authorization review:

1. Deletion or mutation at the SMN1 allele confirmed by genetic testing.
2. Medical records (e.g., chart notes, laboratory values) of the baseline assessment for at least one of the following assessment tools (based on patient age and motor ability) to establish baseline motor ability:
 - i. Hammersmith Infant Neurological Exam Part 2 (HINE-2)
 - ii. Hammersmith Functional Motor Scale Expanded (HFMSSE)
 - iii. Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND)
3. This medication must be prescribed by or in consultation with a physician who specializes in treatment of spinal muscular atrophy.

B. Authorization may be granted for treatment of SMA when all of the following criteria are met:

1. Member has type 1, type 2, or type 3 SMA.
2. There is genetic documentation of 5q SMA homozygous gene mutation, homozygous gene deletion, or compound heterozygote.
3. The member is 25 years of age or younger at initiation of treatment.
4. Member is not dependent on either of the following:
 - i. Invasive ventilation or tracheostomy
 - ii. Use of non-invasive ventilation beyond naps and nighttime sleep



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5. Member meets one of the following criteria:
 - i. Member has not previously received gene replacement therapy for SMA (e.g., Zolgensma), or
 - ii. Member has previously received gene replacement therapy for SMA (e.g., Zolgensma) and has experienced a worsening in clinical status since receiving gene replacement therapy as demonstrated by a decline of minimally clinical important difference from highest score achieved on one of the following exams (based on member age and motor ability):
 - a. HINE-2: Decline of at least 2 points on kicking and 1 point on any other milestone (excluding voluntary grasp)
 - b. HFMSE: Decline of at least 3 points
 - c. CHOP-INTEND: Decline of at least 4 points
6. Member will not use Spinraza and Evrysdi concomitantly.
7. If the member has not received a loading dose, the loading dose will be dosed at 12 mg (5mL) on Day 0, 14, 28, and 58.

Criteria for Continuation of Therapy:

Spinal Muscular Atrophy (SMA)

A. Submission of the following information is necessary for the prior authorization review:

1. This medication must be prescribed by or in consultation with a physician who specializes in treatment of spinal muscular atrophy.
2. Medical records (e.g., chart notes, laboratory values) of the most recent (less than 1 month prior to continuation request) assessment by at least one of the following assessments:
 - i. HINE-2
 - ii. HFMSE
 - iii. CHOP-INTEND
 - iv. For members prescribed Spinraza due to clinical worsening after receiving gene replacement therapy (e.g., Zolgensma): documentation of the impact of Spinraza therapy (e.g., impact on motor milestones).
3. Members who were previously established on Spinraza and subsequently administered gene replacement therapy (e.g., Zolgensma) must meet all initial criteria prior to re-starting therapy on Spinraza.

B. Authorization may be granted for continued treatment of Spinal Muscular Atrophy (SMA) when all of the following criteria are met:

1. Member has type 1, type 2, or type 3 SMA



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2. Member is not dependent on either of the following:
 - I. Invasive ventilation or tracheostomy
 - II. Use of non-invasive ventilation beyond naps and nighttime sleep
3. Submission of medical records (e.g., chart notes, laboratory values) of the most recent (less than 1 month prior to continuation request) assessment documenting a positive clinical response from pretreatment baseline to Spinraza therapy, as demonstrated by at least one of the following assessments:
 - I. HINE-2
 - a. One of the following:
 - i. Member exhibited improvement or maintenance of previous improvement of at least a 2-point (or maximal score) increase in ability to kick; *or*
 - ii. Member exhibited improvement or maintenance of previous improvement of at least a 1-point (or maximal score) increase in any other HINE-2 milestone (e.g., head control, rolling, sitting, crawling, standing, or walking) excluding voluntary grasp; *and*
 - b. One of the following:
 - iii. Member exhibited improvement or maintenance of previous improvement in more HINE-2 motor milestones than worsening (net positive improvement); *or*
 - iv. Member achieved and maintained any new motor milestones when they would otherwise be unexpected to do so (e.g., sit or stand unassisted, walk)
 - II. HFMSE
 - c. One of the following:
 - i. Member exhibited improvement or maintenance of previous improvement of at least a 3-point increase in score; *or*
 - ii. Member has achieved and maintained any new motor milestone from pretreatment baseline when they would otherwise be unexpected to do so
 - III. CHOP-INTEND
 - d. One of the following:



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- i. Member exhibited improvement or maintenance of previous improvement of at least a 4-point increase in score; or
 - ii. Member has achieved and maintained any new motor milestone from pretreatment baseline when they would otherwise be unexpected to do so
4. Member was prescribed Spinraza due to clinical worsening after receiving gene replacement therapy (e.g., Zolgensma) and there is documentation of stabilization or improvement in clinical status with Spinraza therapy (e.g., impact on motor milestones).
5. Member will not use Spinraza and Evrysdi concomitantly
6. If member has already received a loading dose, the maintenance dose will not exceed 12 mg (5 mL) every 4 months

Approval Duration and Quantity Restrictions:

Initial and Renewal Approval: 12 months

References:

1. Spinraza [package insert]. Cambridge, MA: Biogen Inc.; February 2023.
2. Arnold WD, Kassar D, Kissel JT, et al. Spinal muscular atrophy: diagnosis and management in a new therapeutic era. *Muscle & Nerve*. 2015;51(2):157-167.
3. Burgunder JM, Schols L, Baets J, et al. EFNS guidelines for the molecular diagnosis of neurogenetic disorders: motoneuron, peripheral nerve and muscle disorders. *European J Neurol*. 2011;18:207-217.
4. Finkel RS, Chiriboga CA, Vajsar J, et al. Treatment of infantile-onset spinal muscular atrophy with nusinersen: a phase 2, open-label, dose-escalation study. *Lancet*. 2016;388:3017-26.
5. Finkel RS, Mercuri E, Darras BT, et al. Nusinersen versus sham control in infantile-onset spinal muscular atrophy. *N Engl J Med*. 2017; 377:1723-1732.
6. Ionis Pharmaceuticals, Inc. A Study to Assess the Efficacy and Safety of IONIS-SMN Rx in Infants with Spinal Muscular Atrophy. In: ClinicalTrials.gov [internet]. Bethesda (MD): National Library of Medicine (US). 2000- [2016 Feb 14]. Available from: <https://clinicaltrials.gov/ct2/show/NCT02193074>.
7. Mercuri E, Darras BT, Chiriboga CA, et al. Nusinersen versus sham control in later-onset spinal muscular atrophy. *N Engl J Med*. 2018; 378:625-635.
8. Wang CH, Finkel RS, Bertini ES, et al. Consensus statement for standard care in spinal muscular atrophy. *J Child Neurol*. 2007;22(8):1027-1049