MASSACHUSETTS

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Pharmacy Medical Policy

# Spinal Muscular Atrophy (SMA) Medications

## Table of Contents



## Policy Number: 044

BCBSA Reference Number: 5.01.28

## Related Policies

- Quality Care Dosing guidelines may apply to the following medications and can be found in Medical Policy #621A.
- Zolgensma (onasemnogene abeparvovec-xioi) for Spinal Muscular Atrophy (SMA) #008

## Policy

### Commercial Members: Managed Care (HMO and POS), PPO, and Indemnity

**Note:** All requests for outpatient retail pharmacy for indications listed and not listed on the medical policy guidelines may be submitted to BCBSMA Clinical Pharmacy Operations by completing the Prior Authorization Form on the last page of this document. Physicians may also call BCBSMA Pharmacy Operations department at (800)366-7778 to request a prior authorization/formulary exception verbally. Please refer to the chart below for the formulary and/or step status of the medications affected by this policy.

| Standard Formulary | |
|---|---|
| **Drug** | **Formulary Status** |
| Evrysdi ™ | PA Required |
| Spinraza ™ | PA Required |

**We may cover** Evrysdi ™ **(**risdiplam**)** for spinal muscular atrophy (SMA) in patients when **all** of the following criteria are met:

- Diagnosis of spinal muscular atrophy confirmed by genetic testing demonstrating bi-allelic mutations in the survival motor neuron 1 (SMN1) gene as stated below:
  deletion of both copies of the SMN1 gene OR
  identification of pathogenic variant(s) in both copies of the SMN1 gene.

**AND**

- If patient is symptomatic, documentation of a genetic test confirms 2, 3 or 4 copies of the SMN2 gene; OR If patient is asymptomatic, documentation of a genetic test confirms minimum of 2 but less than 4 copies of the SMN2 gene.

**AND**

- The prescription is written by a board certified / board eligible Neurologist.

**AND**

- Patient is not on permanent ventilator dependence

**AND**

- Dose is limited to FDA approved dosing of less than 2 months of age at 0.15mg/kg daily oral dosing or 2 months to less than 2 years of age dosed at 0.2 mg/kg daily oral dosing or for 2 years and older dosed at 0.25 mg/kg with a Max dose of 5mg or 6 & 2/3 mls (20 kg or above) of oral liquid daily.

**AND**

- Patient is not receiving **concurrent** treatment with Spinraza™ (nusinersen) **or** Zolgensma® (onasemnogene abeparvovec).

Reauthorization will require the same criteria above and documentation to support clinically meaningful improvement in motor milestones during previous treatment period.

If approved the Prior Authorization will be granted for up to one year.

**We may cover Spinraza ™ (**nusinersen**)** for spinal muscular atrophy (SMA) in patients when **all** of the following criteria are met:

- Diagnosis of spinal muscular atrophy confirmed by genetic testing demonstrating bi-allelic mutations in the survival motor neuron 1 (SMN1) gene as stated below:
  - deletion of both copies of the SMN1 gene OR
  - compound heterozygous mutations of the SMN1 gene (defined below):
    - pathogenic variant(s) in both copies of the SMN1 gene
    - pathogenic variant in one copy and deletion of the second copy of the SMN1 gene.

**AND**

- If patient is symptomatic, documentation of a genetic test confirms 2, 3 or 4 copies of the SMN2 gene; OR If patient is asymptomatic, documentation of a genetic test confirms minimum of 2 but less than 4 copies of the SMN2 gene.

**AND**

- The prescription is written by a board certified / board eligible Neurologist.

**AND**

- Dose is limited to FDA approved dosing of 12mg (5ml) administered intrathecally per treatment with 4 loading doses; 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.

**AND**

- Patient is not receiving **concurrent** treatment with Evrysdi™ (risdiplam) **or** Zolgensma® (onasemnogene abeparvovec).

Reauthorization will require the same criteria above and documentation to support clinically meaningful improvement in motor milestones during previous treatment period.

If approved the Prior Authorization will be granted for up to one year.

**Requests based exclusively on the use of samples will not meet coverage criteria for exception. Additional clinical information demonstrating medical necessity of the desired medication must be submitted by the requesting prescriber for review.

We do not cover the medications listed above for other conditions not listed above.

## CPT Codes / HCPCS Codes / ICD Codes

*Inclusion or exclusion of a code does not constitute or imply member coverage or provider reimbursement. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage as it applies to an individual member.*

*Providers should report all services using the most up-to-date industry-standard procedure, revenue, and diagnosis codes, including modifiers where applicable.*

**The above medical necessity criteria MUST be met for the following codes to be covered for Commercial Members: Managed Care (HMO and POS), PPO, and Indemnity:**

## HCPCS Codes

| HCPCS codes: | Code Description |
|---|---|
| J2326 | Injection, nusinersen, 0.1 mg |

**The following ICD Diagnosis Codes are considered medically necessary when submitted with the HCPCS code above if medical necessity criteria are met:**

## ICD-10 Diagnosis Codes

| ICD-10-CM diagnosis codes: | Code Description |
|---|---|
| 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 |

**CPT Codes**
There is no specific CPT code for this service.

**Other Information**
Blue Cross Blue Shield of Massachusetts (BCBSMA*) members (other than Medex®; Blue MedicareRx, Medicare Advantage plans that include prescription drug coverage) will be required to fill their prescriptions for the above medications at one of the providers in our retail specialty pharmacy network, see link below:

Link to Specialty Pharmacy List

## Individual Consideration

All our medical policies are written for the majority of people with a given condition. Each policy is based on medical science. For many of our medical policies, each individual's unique clinical circumstances may be considered in light of current scientific literature. Physicians may send relevant clinical information for individual patients for consideration to:

Blue Cross Blue Shield of Massachusetts
Pharmacy Operations Department
25 Technology Place
Hingham, MA 02043
Tel: 1-800-366-7778
Fax: 1-800-583-6289

## Prior Authorization Information

**Outpatient**

For services described in this policy, see below for products where prior authorization **IS REQUIRED** if the procedure is performed **outpatient**.

| | Outpatient |
|---|---|
| **Commercial Managed Care (HMO and POS)** | Prior authorization is **required**. |
| **Commercial PPO** | Prior authorization is **required**. |

## Policy History

| Date | Action |
|---|---|
| 8/2022 | Updated to include new age dosing of Evrysdi ™ to the policy. |
| 12/2021 | BCBSA National medical policy review. No changes to policy statements. New references added. |
| 4/2021 | Updated to align with Association policy with changes in criteria. |
| 10/2020 | Updated to add Evrysdi ™ to the policy. |
| 10/2019 | Updated to reference the Association policy |
| 1/2018 | Clarified coding information |
| 10/2017 | Updated to change Walgreens Specialty Name. |
| 7/2017 | Updated to add AllCare to Pharmacy Specialty list. |
| 5/2017 | Implementation of a new policy including the medication Spinraza™. |

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**To request prior authorization using the Massachusetts Standard Form for Medication Prior Authorization Requests (eForm), click the link below:**
http://www.bluecrossma.org/medical-policies/sites/g/files/csphws2091/files/acquiadam-assets/023%20E%20Form%20medication%20prior%20auth%20instruction%20prn.pdf