MASSACHUSETTS

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Pharmacy Medical Policy

Spinal Muscular Atrophy (SMA) Medications

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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.

Prior Authorization Information

<table>
<tr><td colspan="2">☒ Prior Authorization
☐ Step Therapy
☒ Quality Care Dosing</td><td>Pharmacy Operations:
Tel: 1-800-366-7778
Fax: 1-800-583-6289
Policy last updated | 7/1/2023</td></tr>
<tr><td>Pharmacy (Rx) or Medical (MED) benefit coverage</td><td>☒ Rx
☐ MED</td><td rowspan="2">To request for coverage: Physicians may call, fax, or mail the attached form (Formulary Exception/Prior Authorization form) to the address below.
Blue Cross Blue Shield of Massachusetts
Pharmacy Operations Department
25 Technology Place
Hingham, MA 02043

Individual Consideration: Policy for requests that do not meet clinical criteria of this policy, see section labeled Individual Consideration</td></tr>
<tr><td colspan="2">Policy applies to Commercial Members:
Managed Care (HMO and POS),
PPO and Indemnity
MEDEX with Rx plan
Managed Major Medical with Custom BCBSMA Formulary
Comprehensive Managed Major Medical with Custom BCBSMA Formulary
Managed Blue for Seniors with Custom BCBSMA Formulary</td></tr>
</table>

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 ™ **(**risdiplam**)**	PA Required
Spinraza ™ **(**nusinersen**)**	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

Policy History

Date	Action
7/2023	Reformatted Policy.
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