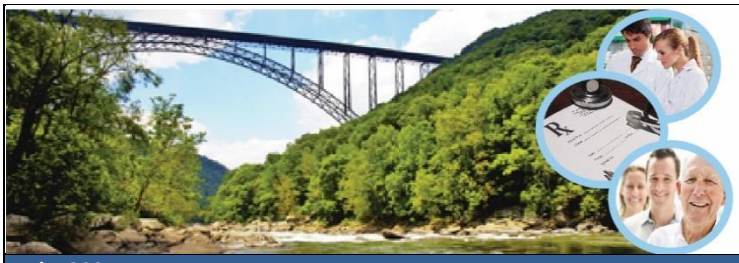

West Virginia Medicaid
Pharmacy Solutions

July, 2021

West Virginia Medicaid Pharmacy Department

https://dhhr.wv.gov/bms/BMS%20Pharmacy

Provider Services

888-483-0793
888-483-0801 (Pharmacy)
304-348-3360
Monday – Friday
8:00 am until 5:00 pm

Pharmacy Help Desk & Pharmacy Prior Authorization (Rational Drug Therapy Program)

800-847-3859 (Phone)
800-531-7787 (Fax)
Monday – Saturday
8:30 am until 9:00 pm
Sunday 12:00 pm until 6:00 pm

Member Services

888-483-0797
304-348-3365
Monday – Friday
8:00 am until 5:00 pm

Preferred Drug List

For a copy of the most recent preferred drug list, visit:
https://dhhr.wv.gov/bms/BMS%20Pharmacy/Pages/Preferred-Drug-List.aspx

State Maximum Allowable Cost (SMAC)

SMAC Review Form:
https://dhhr.wv.gov/bms/BMS%20Pharmacy/SMAC/Pages/default.aspx
Please refer questions to Change Healthcare at 1-855-389-9504 or e-mail to:
PBA_WVSMAC@changehealthcare.com

Spinal Muscular Atrophy Overview

Spinal Muscular Atrophy (SMA) is a group of genetic disorders caused by a mutation in the survival motor neuron 1 (SMN1) gene that regulates the production of the nerves that control the muscles. This causes a degeneration within the spinal cord and lower brainstem resulting in progressive muscle weakness and atrophy which can ultimately be fatal in more severe types. The incidence of SMA ranges from 4 to 10 per 100,000 live births. With 1 in 50 estimated genetic carriers of the mutation within the United States. There are an estimated 2 children born annually with SMA within West Virgnia with and 58 individuals within the state living with SMA.

SMA Types

SMA has various types that differ in onset and symptoms as noted below:

SMA Type 1

Onset: Infantile (before 6 months)
Symptoms: Progressive and rapid. Inability to sit unsupported.
Other: Most common form and the most severe in terms of symptoms and progression. Makes up roughly 60% of cases.

SMA Type 2

Onset; Between 3 and 15 months of age
Symptoms: Less progressive than type 1. Can often sit unassisted but is delayed. No independent standing and walking. Affects legs more than arms.

SMA Type 3

Onset: 18 months and adulthood
Symptoms: Affects legs more than arms. Can stand independent and ambulate independently. However, may progress over time and become wheelchair dependent.

SMA Type 4

Onset: Adult onset
Symptoms: Least severe symptoms of all types.

Diagnosis and Treatment

Diagnosis is often made from a combination of clinical signs and symptoms along with confirmatory genetic testing. Unexplained muscle weakness or low tone, motor difficulties, loss of motor skills, and tongue fasciculations are all signs and symptoms that can be used to initially identify SMA in infants, children, or adults. The diagnosis can then be confirmed by genetic testing.

Traditionally, supportive therapy, such as nutrition, respiratory assistance, orthopedic care, and treating complications of muscle weakness were the mainstays of therapy. Left untreated, most patients with progressive SMA Type 1 will die before 2 years of age. However, more recently disease modifying therapies have become available.

Newer Disease Modifying Therapies

Since 2016 3 newer therapies have been approved that directly modify the disease pathology. These include Spinraza (nusinersen), Zolgensma (onasemnogene), and Evrysdi (risdiplam). Each product is outlined below:

Spinraza (nusinersen)

Mechanisms of Action: Treats SMA caused by mutations in chromosome 5q that lead to SMN protein deficiency. Shown to increase production of full-length SMN proteins.
FDA indication: Treatment of SMA in pediatric and adults patients.
Route: Intrathecal injection
Dosing: 4 loading doses at 14-day intervals then given once every 4 months as maintenance thereafter.

Zolgensma (onasemnogene)

Mechanisms of Action: viral vector-based gene therapy which delivers a copy of the gene encoding the SMN protein thereby increasing SMN protein expression.
FDA indication: Treatment of pediatric patients less than 2 years of age with SMA and a bi-allelic mutation in the SMN1 gene.
Route: IV infusion
Dosing: One time infusion

Evrysdi (risdiplam)

Mechanisms of Action: SMN21 splicing modifier that increases exon 7 inclusion in SMN2 messenger ribonucleic acid (mRNA) and increased production of full-length SMN protein in the brain
FDA indication: Treatment of SMA in patients 2 months of age and older
Route: Oral solution
Dosing: Once daily per body weight and age

Overview and Place in Therapy

While the addition of these disease modifying therapies is a welcome improvement for patients who prior had only supportive therapy, more questions remain. As of the writing of this article there are currently no guidelines that contain all three products and no head-to head comparative trials. There are also no trials or evidence currently showing that a combination of any of the therapies improves outcomes for patients.

The future holds a better understanding of the pathology of SMA, new potential targets for clinical interventions, and a more accurate evaluation of the clinical and cost efficacy of these products over the long-term.

Upcoming PDL Changes

The following changes will be made to the Preferred Drug List (PDL), effective July 1, 2021, pending recommendation and/or approval by the P&T Committee, BMS, and Secretary of DHHR..

For a comprehensive PDL, refer to: https://dhhr.wv.gov/bms/BMS%20Pharmacy/Pages/Preferred-Drug-List.aspx

NEW PREFERRED DRUGS	
THERAPEUTIC CLASS	**RECOMMENDED for PREFERRED STATUS**
HYPOGLYCEMICS, INSULIN AND RELATED AGENTS	Toujeo (insulin glargine)

NEW NON-PREFERRED DRUGS	
THERAPEUTIC CLASS	**RECOMMENDED for NON-PREFERRED STATUS**
ANTIPARKINSONS AGENTS	Ongentys capsules (opicapone)
GLUCOCORTICOIDS, INHALED	Armonair Digihaler (fluticasone)
GLUCOCORTICOIDS, INHALED	Airduo Digihaler (fluticasone/salmeterol)
GUANYLATE CYCLASE STIMULATORS	Verquvo tablets (vericiguat)
HYPOGLYCEMICS, INSULIN AND RELATED AGENTS	Tresiba (insulin degludec)
HYPOGLYCEMICS, INSULIN AND RELATED AGENTS	Semglee (insulin glargine)
IMMUNOSUPPRESSIVES, ORAL	Lupkynis capsule (voclosporin)
LAXATIVES AND CATHARTICS	Sutab tablet (magnesium sulfate, potassium sulfate, sodium sulfate)
MULTIPLE SCLEROSIS AGENTS	Bafiertam capsules (monomethyl fumerate)
MULTIPLE SCLEROSIS AGENTS	Kesimpta injection (ofatumumab)
OPHTHALMICS, ANTI-INFLAMMATORIES-IMMUNOMODULATORS	Eysuvis suspension (lotoprednol)
PAH AGENTS-PROSTACYCLINES	epoprostenol (generic Veletri)
SPINAL MUSCULAR ATROPHY AGENTS	Evrysdi solution (risdiplam)
STEROIDS, TOPICAL	Impeklo lotion (clobetasol propionate)