AvMed

MEDICAL PRIOR AUTHORIZATION/STEP-EDIT REQUEST*

Directions: **The prescribing physician must sign and clearly print name (preprinted stamps not valid) on this request**. All other information may be filled in by office staff; **fax to 1-877-535-1391**. No additional phone calls will be necessary if all information (including phone and fax #s) on this form is correct. **If information provided is not complete, correct, or legible, authorization can be delayed**.

For Medicare Members: Medicare Coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: https://www.cms.gov/medicare-coverage-database/overview-and-quick-search.aspx. Additional indications may be covered at the discretion of the health plan.

Drug Requested: Zolgensma® (onasemnogene abeparvovec-xioi) **IV (Medical) (J-3399)**

MEMBER & PRESCRIBER INFORMATION: Authorization may be delayed if incomplete.

Member Name: _____

Member AvMed #: _____ **Date of Birth:** _____

Prescriber Name: _____

Prescriber Signature: _____ **Date:** _____

Office Contact Name: _____

Phone Number: _____ **Fax Number:** _____

DEA OR NPI #: _____

DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Form/Strength: _____

Dosing Schedule: _____ **Length of Therapy**: _____

Diagnosis: _____ **ICD Code, if applicable:** _____

Weight: _____ **Date:** _____

Coding:

HCPCS	
J3399	Injection, onasemnogene abeparvovec-xioi, per treatment, up to 5x1015 vector genomes [Zolgensma]
ICD-10 Diagnosis	
G12.0	Infantile spinal muscular atrophy, type 1 [Werdnig-Hoffman]
G12.1	Other inherited spinal muscular atrophy

❑ Standard reviews. In checking this box, the timeframe does not jeopardize the life or health of the member or the member's ability to regain maximum function and would not subject the member to severe pain.

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CLINICAL CRITERIA: Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

Zolgensma® is proven and medically necessary for ONE treatment per lifetime for the treatment of spinal muscular atrophy (SMA) in members who meet ALL of the following criteria:

- **Is member currently enrolled in a clinical trial* for an experimental therapy for SMA?**
  ❑ **Yes** ❑ **No**

  If **YES,** chart notes, diagnostics, and lab results documenting experimental therapy trial details must be provided with request**.**

-Drugs are not covered when members are included in a clinical trial. This includes drugs paid for directly by the clinical trial and/or another payor.

- Submission of medical records, chart notes, laboratory values, etc. **confirming** the following:

  ❑ Prescriber must submit baseline documentation of **ONE** of the following **(ACTUAL COMPLETED ASSESSMENT WITH RESULTS MUST BE PROVIDED. DO NOT SEND THE RESULT NUMBER WITHIN THE PROGRESS NOTES.)**
  - Hammersmith Infant Neurological Exam (HINE) (infant to early childhood);

    OR
  - Hammersmith Functional Motor Scale Expanded (HFMSE);

    OR
  - Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND)

    AND

  ❑ Mutation or deletion of genes in chromosome 5q resulting in **ONE** of the following:
  - ❑ Homozygous gene deletion or mutation of SMN1 gene **(e.g., homozygous deletion of exon 7 at locus 5q13)**; **OR**
  - ❑ Compound heterozygous mutation of SMN1 gene (deletion of SMN1 exon 7 [allele 1] and mutation of SMN1 [allele2]);

    AND

  ❑ **ALL** the following have been met:
  - Diagnosis of likely Type 1 SMA based on the results of SMA newborn screening;

    AND

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- Submission of medical records **(e.g., chart notes, laboratory values)** confirming that member has ≤ 2 copies of SMN2 gene;

AND

- Diagnosis of symptomatic SMA by a neurologist with expertise in the diagnosis of SMA;

AND

- For use in a neonatal member born prematurely, the full-term gestational age has been reached (full-term gestational age – 40 weeks)

AND

- Six months of age or younger at the time of vector infusion

OR

- Over 6 months of age at the time of FDA approval on May 24, 2019, but less than 2 years of age at the time of vector infusion

AND

- Previously established on nusinersen (Spinraza®) with a positive and stable clinical response (as evidenced by a Children's Hospital of Philadelphia Infant Test for Neuromuscular Disorders (CHOP INTEND) score of more than 40 points

AND

- Member does not have advanced SMA at baseline (e.g., complete paralysis of limbs, invasive ventilator support (Tracheotomy with positive pressure), requirement of noninvasive ventilator support averaging > 6 hours/day, and anti-AAV9);

AND

- Submission of medical records, chart notes, laboratory values confirming member does not have advanced SMA as defined by the fact that member's most recent CHOP-INTEND score is ≥ 40;

AND

- Member is ≤ 13.5 kg;

AND

- Dose to be administered does not exceed one kit of Zolgensma® 1.1 x 1014 vector genomes (vg) per kg of body weight

AND

❑ Member does not have **either** of the following:

- ❑ Invasive ventilation support (i.e., tracheotomy with positive pressure) or pulse oximetry < 95% saturation);

OR

- ❑ Use of noninvasive ventilator support averaging > 6 hours/day;

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AND

- ❑ Zolgensma® is prescribed by a neurologist with expertise in the treatment of SMA;

AND

- ❑ Member is not to receive routine concomitant SMN modifying therapy (e.g., Spinraza™) **(Member's medical records will be reviewed and any current authorizations for SMN modifying therapy will be terminated upon Zolgensma® approval; member access to subsequent SMN modifying therapy will be assessed according to respective coverage policy of concomitant agent)**;

AND

- ❑ Physician submits the lab assessment for presence of anti-AAV9 antibodies and managed accordingly;

AND

- ❑ Physician attests that the member will not receive Zolgensma if the most recent pre-treatment anti-AAV9 antibody titer is **above** 1:50*;

AND

- ❑ **Physician attests that member, while under the care of the physician, will be assessed by one of the following exam scales during subsequent office visits for a period not to exceed 3 years* ¥**

  - ❑ Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND) scale during subsequent office visits while the member is 2 to 3 years of age or younger***¥**;

  OR

  - ❑ Hammersmith Functional Motor Scale Expanded (HFMSE) during subsequent office visits while the member is 2 to 3 years of age or older;

  OR

  - ❑ Hammersmith Infant Neurological Exam (HINE) (infant to early childhood);

AND

- ❑ Physician would submit to AvMed documentation, not more frequently than bi-annually, of follow-up member assessment(s) including, but not necessarily limited to, serial CHOP INTEND or HFMSE assessments while member is under the care of the physician***¥**;

AND

- ❑ Member will receive prophylactic prednisolone (or glucocorticoid equivalent) prior to and following receipt of Zolgensma® within accordance of the United States Food and Drug Administration (FDA) approved Zolgensma® labeling;

AND

- ❑ Member has never previously received Zolgensma® treatment in their lifetime;

AND

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- ❑ Authorization will be for no longer than 14 days from approval or until 9 months of age, whichever is first;

AND

- ❑ If replacing Spinraza™ (nusinersen), Zolgensma® (onasemnogene) will not be prescribed concurrently as dual therapy or receive concomitant SMN modifying therapy;

AND

- ❑ Absence of the c.859G>C modification in exon 7 of SMN2 gene;

AND

- ❑ Lab values less than 30 days from time of request must be provided with form

***¥ - under the care of the physician**

- ❑ Documentation of baseline laboratory tests of the following to ensure no renal impairment, hepatic impairment or hematologic impairment is present in the following:
  - ❑ Platelet count within normal limits
  - ❑ Troponin-1 within normal limits
  - ❑ Alanine aminotransferase/Aspartate aminotransferase (<2x upper limit of normal)
  - ❑ Total bilirubin within normal limits
  - ❑ Prothrombin time within normal limits
- ❑ Verify member does not have a contraindication or intolerant to corticosteroid therapy

  (Concomitant therapy will be implemented with systemic corticosteroids equivalent to oral prednisolone 1mg/kg/day for 30 days, starting one day prior to administration of Zolgensma®)

Medication being provided by (check box below that applies):

- ❑ **Location/site of drug administration:** _____

  NPI or DEA # of administering location: _____

OR

- ❑ **Specialty Pharmacy - PropriumRx**

For urgent reviews: Practitioner should call AvMed Pre-Authorization Department if they believe a standard review would subject the member to adverse health consequences. AvMed's definition of urgent is a lack of treatment that could seriously jeopardize the life or health of the member or the member's ability to regain maximum function.

*****Use of samples to initiate therapy does not meet step edit/ preauthorization criteria.*****

****Previous therapies will be verified through pharmacy paid claims or submitted chart notes.****

***Approved by Pharmacy and Therapeutics Committee:** 7/18/2019
REVISED/UPDATED: ~~9/25/2019; 12/4/2020;~~ **10/21/2021**