zolgensma®
(onasemnogene abeparvovec-xioi)
suspension for intravenous infusion

Payer Coding and Billing Guide

Please see Indication and Important Safety Information on page 3 and click here for Full Prescribing Information, including Boxed WARNING for Serious Liver Injury and Acute Liver Failure.

An overview of this guide

This guide has been developed to provide an overview of applicable coding and billing information for ZOLGENSMA® (onasemnogene abeparvovec-xioi) where health plans provide coverage for ZOLGENSMA as part of the medical benefit. ZOLGENSMA is available to purchase from CuraScript SD® for Buy and Bill treatment centers and through a limited network of Specialty Pharmacies.

ZOLGENSMA Distribution

BUY AND BILL

Ordered from
CuraScript SD®
Phone: **866-263-8464**

LIMITED NETWORK SPECIALTY PHARMACIES

Novartis Gene Therapies has partnered with select Specialty Pharmacies to support ZOLGENSMA.

This guide provides codes* for

Product ordering through both Buy and Bill and Specialty Pharmacy	**Infusion codes for ZOLGENSMA†**	**SMA diagnosis**

*Codes provided in this guide may be subject to change. Codes are provided in this guide for illustrative purposes only. It is the health care provider's responsibility to choose the code or codes appropriate for each patient.

†Codes related to other aspects of spinal muscular atrophy (SMA) or its comorbidities are not included.

Indication and Important Safety Information

zolgensma® (onasemnogene abeparvovec-xioi) suspension for intravenous infusion

INDICATION

ZOLGENSMA is an adeno-associated virus (AAV) vector-based gene therapy indicated for the treatment of pediatric patients less than 2 years of age with spinal muscular atrophy (SMA) with bi-allelic mutations in the *survival motor neuron 1* (*SMN1*) gene.

Limitations of Use

The safety and effectiveness of repeat administration or the use in patients with advanced SMA (eg, complete paralysis of limbs, permanent ventilator dependence) has not been evaluated with ZOLGENSMA.

IMPORTANT SAFETY INFORMATION

BOXED WARNING: Serious Liver Injury and Acute Liver Failure

Cases of acute liver failure with fatal outcomes have been reported. Acute serious liver injury, acute liver failure, and elevated aminotransferases can also occur with ZOLGENSMA. Patients with preexisting liver impairment may be at higher risk. Prior to infusion, assess liver function of all patients by clinical examination and laboratory testing. Administer systemic corticosteroid to all patients before and after ZOLGENSMA infusion. Continue to monitor liver function for at least 3 months after infusion, and at other times as clinically indicated. If acute serious liver injury or acute liver failure is suspected, promptly consult a pediatric gastroenterologist or hepatologist.

WARNINGS AND PRECAUTIONS

Systemic Immune Response

Patients with underlying active infection, either acute or chronic uncontrolled, could be at an increased risk of serious systemic immune response. Administer ZOLGENSMA to patients who are clinically stable in their overall health status (eg, hydration and nutritional status, absence of infection). Postpone ZOLGENSMA in patients with infections until the infection has resolved and the patient is clinically stable.

Thrombocytopenia

Transient decreases in platelet counts, some of which met the criteria for thrombocytopenia, were typically observed within the first 2 weeks after ZOLGENSMA infusion. Monitor platelet counts before ZOLGENSMA infusion and on a regular basis for at least 3 months afterwards.

Thrombotic Microangiopathy

Cases of thrombotic microangiopathy (TMA) were reported to occur generally within the first 2 weeks after ZOLGENSMA infusion. TMA can result in life-threatening or fatal outcomes. Obtain baseline creatinine and complete blood count before ZOLGENSMA infusion. Following infusion, monitor platelet counts closely as well as other signs and symptoms of TMA. Consult a pediatric hematologist and/or pediatric nephrologist immediately to manage as clinically indicated.

Elevated Troponin-I

Increases in cardiac troponin-I levels were observed following ZOLGENSMA infusion. Monitor troponin-I before ZOLGENSMA infusion and on a regular basis for at least 3 months afterwards. Consider consultation with a cardiologist if troponin elevations are accompanied by clinical signs or symptoms.

AAV Vector Integration and Risk of Tumorigenicity

There is a theoretical risk of tumorigenicity due to integration of AAV vector DNA into the genome. Cases of tumor have been reported in patients who received ZOLGENSMA post-approval; a causal relationship has not been established based on tumor analysis. In some cases, limited information was available. Report cases of tumor development in patients who received ZOLGENSMA to Novartis Gene Therapies, Inc. at 1-833-828-3947.

ADVERSE REACTIONS

The most commonly observed adverse reactions (incidence ≥5%) in clinical studies were elevated aminotransferases and vomiting.

Please click here for Full Prescribing Information.

Dosing and NDC designation

The recommended dose of ZOLGENSMA® (onasemnogene abeparvovec-xioi) for single-dose intravenous (IV) infusion in pediatric patients less than 2 years of age is 1.1×10^{14} vector genomes (vg)/kg. The ZOLGENSMA kit consists of 2 to 14 vials, provided in 2 fill volumes (either 5.5 mL or 8.3 mL), each with a nominal concentration of 2.0×10^{13} vg/mL.[1]

The appropriate ZOLGENSMA dose and kit is determined by patient body weight. The table below lists 37 ZOLGENSMA kits appropriate for dosing patients weighing between 2.6 kg and 21.0 kg.[1]

ZOLGENSMA Kit Sizes

PATIENT WEIGHT (kg)	DOSE VOLUME[a] (mL)	KIT CONFIGURATION			NDC NUMBER
		5.5 mL VIALS[b]	8.3 mL VIALS[c]	No. OF VIALS IN KIT	
2.6 – 3.0	16.5	0	2	2	71894-120-02
3.1 – 3.5	19.3	2	1	3	71894-121-03
3.6 – 4.0	22.0	1	2	3	71894-122-03
4.1 – 4.5	24.8	0	3	3	71894-123-03
4.6 – 5.0	27.5	2	2	4	71894-124-04
5.1 – 5.5	30.3	1	3	4	71894-125-04
5.6 – 6.0	33.0	0	4	4	71894-126-04
6.1 – 6.5	35.8	2	3	5	71894-127-05
6.6 – 7.0	38.5	1	4	5	71894-128-05
7.1 – 7.5	41.3	0	5	5	71894-129-05
7.6 – 8.0	44.0	2	4	6	71894-130-06
8.1 – 8.5	46.8	1	5	6	71894-131-06
8.6 – 9.0	49.5	0	6	6	71894-132-06
9.1 – 9.5	52.3	2	5	7	71894-133-07
9.6 – 10.0	55.0	1	6	7	71894-134-07
10.1 – 10.5	57.8	0	7	7	71894-135-07
10.6 – 11.0	60.5	2	6	8	71894-136-08
11.1 – 11.5	63.3	1	7	8	71894-137-08
11.6 – 12.0	66.0	0	8	8	71894-138-08
12.1 – 12.5	68.8	2	7	9	71894-139-09
12.6 – 13.0	71.5	1	8	9	71894-140-09
13.1 – 13.5	74.3	0	9	9	71894-141-09
13.6 – 14.0	77.0	2	8	10	71894-142-10
14.1 – 14.5	79.8	1	9	10	71894-143-10
14.6 – 15.0	82.5	0	10	10	71894-144-10
15.1 – 15.5	85.3	2	9	11	71894-145-11
15.6 – 16.0	88.0	1	10	11	71894-146-11
16.1 – 16.5	90.8	0	11	11	71894-147-11
16.6 – 17.0	93.5	2	10	12	71894-148-12
17.1 – 17.5	96.3	1	11	12	71894-149-12
17.6 – 18.0	99.0	0	12	12	71894-150-12
18.1 – 18.5	101.8	2	11	13	71894-151-13
18.6 – 19.0	104.5	1	12	13	71894-152-13
19.1 – 19.5	107.3	0	13	13	71894-153-13
19.6 – 20.0	110.0	2	12	14	71894-154-14
20.1 – 20.5	112.8	1	13	14	71894-155-14
20.6 – 21.0	115.5	0	14	14	71894-156-14

kg, kilogram; mL, milliliter; NDC, National Drug Code.

[a]Dose volume is calculated using the upper limit of the patient weight range for pediatric patients less than 2 years of age between 2.6 kg and 21.0 kg.

[b] Vial nominal concentration is 2.0×10^{13} vg/mL and contains an extractable volume of not less than 5.5 mL.

[c] Vial nominal concentration is 2.0×10^{13} vg/mL and contains an extractable volume of not less than 8.3 mL.

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HCPCS code

ZOLGENSMA® (onasemnogene abeparvovec-xioi) is billed under an HCPCS J-code. Effective July 1, 2020, the Centers for Medicare & Medicaid Services assigned ZOLGENSMA the following permanent J-code: J3399.[2]

HCPCS code for ZOLGENSMA[3]

DESCRIPTION	CODE
Injection, onasemnogene abeparvovec-xioi, per treatment, up to 5×10^{15} vector genomes	**J3399**

HCPCS, Healthcare Common Procedure Coding System.

CPT codes

ZOLGENSMA® (onasemnogene abeparvovec-xioi) is administered via a single-dose IV infusion.[1] The following Current Procedural Terminology (CPT) codes may be used by health care providers and their staff when coding and billing for ZOLGENSMA infusion. **Please note that these codes do not include office visits for diagnosis and prescribing of medication.**

CPT codes for ZOLGENSMA infusion[4]

DESCRIPTION	CODE
IV infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to one hour	96365
Initial hospital inpatient or observation care, per day, for the evaluation and management of a patient, which requires a medically appropriate history and/or examination and straightforward or low level, moderate level, or high level of medical decision making	99221-99223
Hospital inpatient or observation care, for the evaluation and management of a patient including admission and discharge on the same date, which requires a medically appropriate history and/or examination and straightforward or low level, moderate level, or high level of medical decision making	99234-99236

ICD-10 codes

Please refer to the following code for a patient's diagnosis.

ICD-10 codes for SMA[5]

CONDITION	CODE
Infantile SMA, Type 1 [Werdnig-Hoffman]	G12.0
Other inherited SMA • Adult form SMA • Childhood form, type 2 SMA • Distal SMA • Juvenile form, type 3 SMA [Kugelberg-Welander] • Progressive bulbar palsy of childhood [Fazio-Londe] • Scapuloperoneal form SMA	G12.1
Other SMAs and related syndromes	G12.8
SMA, unspecified	G12.9

ICD-10, International Classification of Diseases, 10th Revision.

*Please contact your **Director of National Accounts** if you have any questions regarding ZOLGENSMA® (onasemnogene abeparvovec-xioi).*

References: 1. ZOLGENSMA. Prescribing information. Novartis Gene Therapies, Inc. **2.** HCPCS release & code sets. Centers for Medicare & Medicaid Services. Accessed November 27, 2024. https://www.hhs.gov/guidance/document/hcpcs-release-code-sets **3.** HCPCS quarterly update. Centers for Medicare & Medicaid Services website. Accessed November 27, 2024. https://www.cms.gov/medicare/coding-billing/healthcare-common-procedure-system/quarterly-update **4.** American Medical Association. CPT® Professional 2023 Edition. Chicago, IL: American Medical Association; 2023. **5.** ICD-10-CM tabular list of diseases and injuries. Centers for Medicare & Medicaid Services website. Accessed November 27, 2024. https://www.cms.gov/medicare/coding-billing/icd-10-codes

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