



State of California—Health and Human Services Agency
Department of Health Care Services



GAVIN NEWSOM
GOVERNOR

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TO: All County California Children Services Program Administrators, Medical Consultants, and Integrated Systems of Care Division Staff

SUBJECT: Treatment for Spinal Muscular Atrophy

I. PURPOSE

The purpose of this Numbered Letter (N.L.) is to update the California Children's Services (CCS) Program's policy regarding the authorization of treatment for 5q spinal muscular atrophy (SMA) for CCS Program clients with confirmed 5q SMA who have bi-allelic mutations in the survival motor neuron 1 (SMN1) gene.

The CCS Program publishes this N.L. under the program's authority to authorize services that are medically necessary to treat CCS-eligible conditions.^{1,2,3}

II. BACKGROUND

5q SMA is an autosomal recessive neurodegenerative disorder, characterized by progressive muscle atrophy and weakness. The weakness is caused by absence or very low levels of survival motor neuron (SMN) protein with resulting degeneration of anterior horn cells in the spinal cord and motor neurons in cranial nerve nuclei of the lower brainstem. The condition is the most common genetic cause of childhood mortality, with an incidence of approximately 1 in 10,000 live births or about 500 new SMA cases per year.

5q SMA results in a continuous spectrum of clinical severity and is classified into four types based on age of onset and severity. SMA1, also known as infantile SMA, is the most common and severe type, affecting approximately 60 percent of all individuals with 5q SMA. Signs appear before six months of age in SMA1; affected individuals never sit, and the majority do not survive past two years of age. SMA2 is clinically evident later in infancy and affected individuals achieve the ability to sit but never walk; individuals with SMA2 typically survive into adulthood if provided with optimal supportive care. SMA3 comes to medical attention in childhood and SMA4 in

adulthood. Individuals with SMA3 and SMA4 have near-normal lifespans and achieve the ability to walk, though they may lose that function as the condition progresses. The motor deficits continuously worsen in all forms of SMA, so improvement or stabilization of motor function are inconsistent with the natural history of the disease.

5q SMA is caused by the deficiency of the survival motor neuron (SMN) protein, which is essential for ribonucleic acid (RNA) processing of gene products required for motor neuron survival. SMN protein is produced by two genes, SMN1 and SMN2, both on chromosome 5q. In unaffected individuals, most SMN protein is the product of SMN1, with only a small amount from the back-up gene, SMN2. SMN1 is transcribed and translated to the fully functional full length SMN protein; SMN2, with a single nucleotide change, produces mostly truncated unstable protein with a approximately 5-10 percent of SMN2 transcripts producing full length functional SMN protein.

5q SMA is inherited as a recessive disorder; all affected individuals are completely missing functional copies of SMN1, and depend on SMN2 for all SMN protein. Worldwide, 1 in 35-50 people have one chromosome 5 on which SMN1 has been deleted without producing any motor neuron abnormalities. Most individuals with 5q SMA have an inherited deletion of SMN1 from each parent. Approximately five percent of 5q SMA patients have inherited one allele with a deletion of SMN1 and one allele with a nonsense or other null mutation of SMN1. In ten percent of patients, one parent is not a carrier of any SMN1 mutation, but the affected offspring has a de novo mutation.

Individuals with 5q SMA have no functional SMN1 genes and rely on their copies of the SMN2 gene for all SMN protein production, which results in low levels of the SMN protein. The severity of disease varies between individuals in large part because of differences in SMN2 copy number. Individuals with SMA who have more copies of SMN2 genes generally have less severe forms of SMA as follows:

1. An individual with a single SMN2 gene and no copies of SMN1 is severely affected at birth;
2. Infants with SMA type 1 generally have two copies of SMN2;
3. Children with type 2 usually have three copies of SMN2 and;
4. Those with SMA type 3 or SMA type 4 have more than 3 copies.

Although SMN protein is ubiquitously expressed, its absence in 5q SMA primarily affects motor neurons, though lesser effects have been documented in other tissues.

Advances in nutrition and pulmonary care for SMA patients have increased survivability. Without treatment targeting the mechanism of the disease, however, patients with SMA experience steady loss of motor function and early death.

Nusinersen (Spinraza) was the first Food and Drug Administration (FDA) approved disease-modifying treatment for 5q SMA in December 2016. Nusinersen is an antisense oligonucleotide that modulates the splicing of SMN2 gene, leading to a higher percentage of transcripts containing exon 7, and more functional SMN protein. Since all patients with 5q SMA carry and rely on their copies of SMN2 for production of SMN protein, all 5q SMA patients are treatable by nusinersen. Consequently, the FDA approved the drug for individuals of all ages with all types of 5q SMA.

Studies to date indicate that the vast majority of patients treated with nusinersen have a positive response, with significant improvement or slower decline in disease progression. The time to response varies significantly, with some patients responding up to 12 months after initial treatment. Presymptomatic infantile onset SMA patients treated with nusinersen had higher achievement of motor milestones than symptomatic infants did in a concurrent trial, suggesting the greatest benefit with early treatment. While nusinersen is effective for many clients, it requires intrathecal delivery and initial loading doses and then continuing administration every four months. It effectively puts a band-aid on a defective gene.⁴

A new gene therapy, onasemnogene abeparvovec (Zolgensma), offers a longer-lasting treatment. Onasemnogene abeparvovec was approved by the FDA on May 24, 2019, for children with SMA aged less than two years with bi-allelic mutations in the SMN1 gene. This treatment uses a single intravenous infusion of an adeno-associated virus serotype 9 vector (AAV9) designed to deliver a functional copy of the gene encoding the SMN protein, replacing the defective or deficient SMN1 gene.

Early results indicate that onasemnogene abeparvovec is associated with reduced need for ventilator support and improved motor function, but there is no evidence that onasemnogene abeparvovec reverses pre-existing nerve damage. Because acute serious liver injury and elevated transaminases can occur with onasemnogene, liver function testing prior to infusion and for at least three months after infusion is recommended.

On August 7, 2020, a new oral formulation was approved to treat SMA for patients aged two months and over. Risdiplam (Evrysdi) is the first oral form to be approved. In vitro assays and animal studies have shown the drug to increase exon 7 inclusion in SMN2 mRNA, leading to increases in SMN protein.

While disease modifying treatment is a critical part of SMA care, it is vital that other supportive care provided through the Special Care Center (SCC), including

pulmonary services, nutrition support, and orthopedic and rehabilitation services are aggressively continued concurrently with nusinersen and risdiplam and following onasemnogene abeparvovec administration.

III. POLICY

Effective the date of this letter, the CCS Program may authorize treatment for 5q SMA as follows:

A. Nusinersen (Spinraza) and Risdiplam (Evrysdi)

1. Eligibility

Nusinersen and risdiplam are benefits for CCS Program clients when all the following criteria are met:

- a. Genetic testing results demonstrate homozygous SMN1 deletion, or any combination of SMN1 deletions or other mutations that result in the functional loss of all SMN1 genes.
- b. In addition to demonstrating loss of functional SMN1 genes, genetic test results includes the number of copies of SMN2.
- c. The client is under the care of one of the following CCS Program approved center types: Neuromuscular Medicine SCC, Neuromusculoskeletal SCC, or Pediatric Rehabilitation SCC.
- d. The client has either of the following:
 - (1) Pre-symptomatic: Defined by genetic testing demonstrating a homozygous SMN1 deletion or mutation, and ≤ 3 copies of SMN2.
 - (2) Symptomatic: Patient with clinical signs of SMA with level of function necessary to preserve communication, for instance finger or eye movements in response to prompt by examiner.
- e. For nusinersen, it can be safely administered intrathecally (IT), taking into consideration the client's scoliosis status. Specifically, for older clients with SMA, the drug may only be authorized if beneficiary has any of the following:
 - (1) No scoliosis.
 - (2) Scoliosis without spine surgery.

- (3) Scoliosis post spine surgery with preserved window of accessibility for intrathecal injection, under fluoroscopic or ultrasound guidance if needed.
 - (4) Scoliosis post spine surgery (i.e. fusion) but with surgical placement of an indwelling catheter or establishment of a new window for IT accessibility.
- f. The client does not have a coexisting terminal condition or a condition with which the risk of nusinersen treatment outweighs the potential benefit.

2. Authorization

- a. For initial authorizations, a CCS Program approved Rehabilitation, Neuromuscular, or Neuromusculoskeletal SCC should submit the following:
 - (1) Medical note from neuromuscular specialist at the SCC containing:
 - (a) Patient demographics, including age of onset.
 - (b) Results of genetic testing, including name of laboratory, number of copies of SMN2, and whether SMN1 sequencing was done.
 - (c) Neurologic status, specifically if client is non-sitter, sitter or walker.
 - (d) Pulmonary status (for example hours of ventilation or Bilevel Positive Airway Pressure [BiPAP]),
 - (e) Nutrition and dietary status (with review by registered dietitian)
 - (f) Results of at least one neuromotor assessment with a score, performed by or under the direction of the authorized SCC, used to establish a clinical baseline. The following are suggested, but any validated assessment may be used at baseline and repeated annually.
 - (i) For non-sitters:
 - Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND) or,⁵
 - Hammersmith Infant Neurological Exam-Part 2 (HINE-2).

- (ii) For sitters:
 - Hammersmith Functional Motor Scale, Expanded (HF MSE) or,
 - Revised Upper Limb Module (RULM).
 - (iii) For walkers:
 - The Timed up and Go test (TUG),
 - The 6 minute walk test or,
 - The 10-meter run/walk test.
 - (iv) For non-ambulatory older clients:
 - Revised Upper Limb Module (RULM),
 - Standard muscle strength assessment.
- (2) Copy of nusinersen or risdiplam prescription by CCS Program paneled neurologist or physical medicine and rehabilitation specialist at the SCC where the client completed evaluation for nusinersen.
- (3) Genetic laboratory confirmation of diagnosis.
- (4) Client has not received onasemnogene abeparvovec.
- b. The CCS Program may reauthorize nusinersin or risdiplam treatment if a CCS-approved Rehabilitation, Neuromuscular or Neuromusculoskeletal SCC has submitted the following documentation to the independent county CCS Program or to Integrated Systems of Care Division (ISCD):
- (1) Date of initial nusinersen or risdiplam treatment.
 - (2) SCC progress notes documented within six months of the authorization request, including a specific description of changes in neuromotor status since initiation of medication, and any drug-related toxicity.
 - (3) Copy of nusinersen or risdiplam prescription by CCS Program paneled neurologist or physical medicine and rehabilitation specialist, or designee, at SCC where evaluation was completed.

- (4) Neuromotor assessment, completed at the SCC within 12 months of the reauthorization request, which demonstrates improvement or lack of deterioration since initiation of nusinersen or risdiplam, with positive response to medication documented by comparing scores to the results prior to medication.
- c. The request is for the FDA-approved dosage only, with the approved loading and maintenance schedules.
 - (1) Nusinersen is a 12 mg suspension, to be administered intrathecally.
 - (2) The nusinersen treatment schedule consists of four loading doses of 12mg, at days 1, 15, 29, and 59, and maintenance doses every 4 months thereafter.

B. Onasemnogene abeparvec (Zolgensma)

1. Eligibility

Onasemnogene abeparvec is a benefit for CCS Program clients when all the following criteria are met:

- a. The client is under the age of two years.
- b. Client has bi-allelic mutations in SMN1 gene, demonstrated by genetic testing results with documentation of both of the following:
 - (1) Genetic documentation of bi-allelic mutations in the SMN1 gene (deletions or point mutations).
 - (2) Documentation of up to and including four copies of SMN2.
- c. Client does not have advanced SMA, as evidenced by any of the following:
 - (1) Invasive ventilator support (tracheostomy with ventilator).
 - (2) Complete paralysis of limbs.
- d. The client is under the care of an approved Neuromuscular SCC, Neuromusculoskeletal SCC, or Pediatric Rehabilitation SCC.
- e. Client does not have AAV9 titer >1:50 as determined by Enzyme-Linked Immunosorbent Assay (ELISA) binding immunoassay.

- f. There is no indication of significant liver injury.
 - g. Client is not currently being treated with nusinersen or risdiplam, or treatment will be discontinued prior to the administration of onasemnogene abeparvovec.
 - h. Client was not previously treated with onasemnogene abeparvovec. Onasemnogene abeparvovec is a one-time treatment, and shall be authorized only once per client.
2. Authorization:

Providers requesting authorization of onasemnogene abeparvovec must provide the following documentation:

- a. Copy of onasemnogene abeparvovec prescription by CCS Program panned neurologist or physical medicine and rehabilitation specialist at the SCC where evaluation for onasemnogene abeparvovec was completed.
 - b. Medical documentation of SCC visit with history and physical examination, including description of plan for onasemnogene abeparvovec administration.
 - c. Genetic laboratory confirmation of diagnosis and number of SMN2 copies.
 - d. Documentation of AAV9 titer that is less than 1:50, within 90 days of planned administration.
 - e. At least one neuromotor assessment, performed within 12 months of the authorization request, with a score used to establish a clinical baseline.
 - f. Documentation of baseline liver function test, platelet counts, and troponin-I.
- C. Additional considerations for medical necessity determination:

For clients who do not meet the criteria described in sections III.A.1., III.A.2., III.B.1., and III.B.2., requesting SCCs may demonstrate medical necessity by submitting any other clinical documentation and/or evidence that would support the initial or reauthorization of the client's treatment for 5q SMA. SCCs should submit this documentation to the ISCD Medical Director or designee.

- D. Whole Child Model (WCM) counties:

For CCS clients who are enrolled in a Medi-Cal managed care plan (MCP) and reside in a WCM county, the client's MCP shall be responsible for authorizing, coordinating, and covering the client's treatment of 5q SMA. MCPs operating in WCM counties should use the authorization guidelines described in this N.L., or utilize the MCP's existing 5q SMA treatment policies, whichever is less restrictive.

IV. POLICY IMPLEMENTATION

A. Nusinersen (Spinraza) and Risdiplam (Evrysdi)

1. Nusinersen and risdiplam are not covered by a Service Code Grouping (SCG) authorization.⁶ SCCs or pharmacies should submit a separate Service Authorization Request (SAR) and supporting documentation in the following manner:
 - a. For nusinersen outpatient administration, as a Hospital or Physician-Administered Drug (PAD):
 - (1) Dates of service beginning January 1, 2018, use Healthcare Common Procedure Coding system (HCPCS) code, J2326. One unit of J2326 = injection, nusinersen, 0.1mg.
 - (2) SCG02 or SCG01 with additional codes needed for procedures and equipment related to nusinersen administration.
 - b. For pharmacy dispensing nusinersen, when the drug is dispensed by a pharmacy provider and delivered to the provider administering the drug:

Authorize its National Drug Code (NDC) to pharmacy.
 - c. For risdiplam, authorize its NDC to the dispensing pharmacy. Currently, the commercial product is available as a 60mg bottle, reconstituted by the pharmacy to provide 80 mls of an oral solution with a concentration of 0.75mg/ml.
2. Requesting CCS Program providers must submit the following items to their clients' local CCS Program county office for clients who live in independent counties, or directly to the ISCD Special Populations Authorization Unit for clients who live in dependent CCS counties:
 - a. CCS Program SAR

- b. Medical documentation from the CCS Program approved SCC, with neuromotor assessment scores every 12 months and summary of changes in neuromotor status every six months.
 - c. Supporting documentation described in III.A.2.(a) or III.A.2.(b) above.
3. When the County CCS Program determines that the request and documentation submitted by the SCC is complete, the county will send a SAR and forward the request) and supporting documentation to: CCSExpeditedReview@dhcs.ca.gov or via secure Right fax number: (916) 440-5306.
 4. The State CCS Program office will issue the authorization.
 5. The State CCS Program office will issue initial authorization for a period of twelve months or until the end of program eligibility period.
 6. Reauthorization shall be granted every twelve months following review of documentation described above unless there are significant adverse effects or change in eligibility.
 7. Reauthorizations will be done by the independent county CCS Program or ISCD Special Populations Authorization Unit for dependent counties.

B. Onasemnogene abeparvovec (Zolgensma)

1. Onasemnogene abeparvovec is not covered by a SCG authorization⁶ and a separate authorization is needed for outpatient administration.
2. Requesting CCS Program providers must submit the following items to their beneficiaries' local CCS Program county office or ISCD Special Populations Authorization Unit:
 - a. CCS Program SAR with Outpatient National Provider Identifier number for:
 - (1) HCPCS code, J3590 on one service line, for dates of service up to June 30, 2020. Beginning July 1, 2020, J3399 must be used.
J3399 = Injection, onasemnogene abeparvovec-xioi, per treatment, up to 5×10^{15} vector genomes
 - (2) Supporting clinical documentation should justify medical necessity and that the service is the least-costly to meet the client's needs.
 - (3) Request Unit = 3 for J3590 or J3399.

- (4) SCG02 or SCG01 with additional codes needed for procedures and equipment related to onasemnogene abeparvovec administration.
3. When the County CCS Program determines that the request and documentation submitted by the SCC is complete, the county will send a SAR and forward the request and supporting documentation to:
CCSExpeditedReview@dhcs.ca.gov or via secure Right fax number: (916) 440-5306.
4. The State CCS Program office will issue the authorization.
5. Each CCS client is eligible to receive only one treatment of Onasemnogene Abearvovec, under J3590, J3399, or any other code (HCPCS, Current Procedural Terminology [CPT], or by NDC).
6. Requesting providers must adhere to the following special instructions when filing a claim:
 - a. Submit three (3) claim lines, each claim for one (1) unit on UB-04 paper claim form or 837I (Institutional) electronic claim form;
 - b. Claims submitted with one or two claim lines, will be denied
 - c. Each claim line can accommodate up to \$999,999 billed amount;
 - d. Total reimbursement of the three claim lines will be no more than the invoice price paid by provider.
 - e. Providers must identify Zolgensma paper claims by notation as such in the remarks section of the paper claim. For electronic claims, provider shall indicate claim is for Zolgensma on a coversheet, to ensure that these are processed expeditiously.
 - f. Providers should note that except for the first claim line, payment for any additional line will be delayed for 2-3 additional weeks due to systems constraints.
 - g. Payment for Zolgensma shall be a once in a lifetime reimbursement under J3590, J3399, or any other code (HCPCS, CPT, or by NDC).

Beginning April 1, 2021, all requests for prior authorization of medications billed by NDC and dispensed by a Medi-Cal enrolled pharmacy provider, shall be sent from the

pharmacy provider to the Medi-Cal Rx vendor, Magellan Medicaid Administration, Inc. (Magellan). The Medi-Cal RX website provides guidance: <https://medi-calrx.dhcs.ca.gov/home/>.

If you have any questions regarding this N.L., please contact the ISCD Medical Director or designee, via e-mail at ISCD-MedicalPolicy@dhcs.ca.gov

Sincerely,

ORIGINAL SIGNED BY

Roy Schutzengel
Medical Director
Integrated Systems of Care Division

Attachment(s):

Attachment 1: Current Treatment Options for Spinal Muscular Atrophy

Attachment 2: Notice to Providers Regarding Special Billing

¹ 22 Cal. Code Regs. § 41515.1 et. seq. Determination of Medical Eligibility
<https://govt.westlaw.com/calregs/Document/I28E30090D4B811DE8879F88E8B0DAAAE?viewType=FullText&originContext=documenttoc&transitionType=CategoryPageItem&contextData=%28sc.Default%29>

² 22 Cal. Code Regs. § 41700 Availability
[https://govt.westlaw.com/calregs/Document/I2F1A7E70D4B811DE8879F88E8B0DAAAE?viewType=FullText&originContext=documenttoc&transitionType=CategoryPageItem&contextData=\(sc.Default\)&bhcp=1&ignorebhwarn=ignoreWarns](https://govt.westlaw.com/calregs/Document/I2F1A7E70D4B811DE8879F88E8B0DAAAE?viewType=FullText&originContext=documenttoc&transitionType=CategoryPageItem&contextData=(sc.Default)&bhcp=1&ignorebhwarn=ignoreWarns)

³ 22 Cal. Code Regs. § 41740 Eligibility for Treatment Services
<https://govt.westlaw.com/calregs/Document/I2FDD8050D4B811DE8879F88E8B0DAAAE?viewType=FullText&originContext=documenttoc&transitionType=StatuteNavigator&contextData=%28sc.Default%29>

⁴ Al-Zaidy S, Pickard AS, Kotha K, et al. Health outcomes in spinal muscular atrophy type 1 following AVXS-101 gene replacement therapy. *Pediatr Pulmonol.* 2019;54(2):179-185
<https://www.ncbi.nlm.nih.gov/pubmed/30548438>

⁵ Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND)
<https://www.ncbi.nlm.nih.gov/pubmed/20074952>

⁶ Service Authorization Request Tools
<https://www.dhcs.ca.gov/services/ccs/cmsnet/Pages/SARTools.aspx#service>

Current Treatment Options for Spinal Muscular Atrophy

Generic Name	Nusinersen	Onasemnogene abeparvovec	Risdiplam
Brand Name	Spinraza	Zolgensma	Evrysdi
FDA Indication	Treatment of spinal muscular atrophy (SMA) in pediatric and adult patients	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.	Treatment of spinal muscular atrophy (SMA) in patients 2 months of age and older.
Mechanism of Action	Survival motor neuron-2 (SMN2)-directed antisense oligonucleotide	Adeno-associated virus vector-based gene therapy	Survival of motor neuron 2 (SMN2) splicing modifier
FDA approved age	All ages	Less than 2 years of age	Two months of age and older
Dosing	The recommended dosage is 12 mg (5 mL) per administration	Recommended dosage of ZOLGENSMA is 1.1×10^{14} vector genomes (vg) per kg of body weight.	For 2 months to < 2 years of age: 0.2mg/kg daily dose For 2 years of age and older weighing < 20kg: 0.25mg/kg daily dose For 2 years of age and older and weighing 20kg or more: 5mg daily dose
Dosing Regimen	Initiate nusinersen 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	Single-dose intravenous infusion only	Daily dose after a meal using provided oral syringes

Department of Health Care Services (DHCS)**Pharmacy Benefit Division****Pharmacy Policy Branch****NOTICE TO PROVIDERS REGARDING THE SPECIAL BILLING OF ZOLGENSMA CLAIMS EFFECTIVE JULY 1, 2020.**

The Department of Health Care Services (DHCS) would like to notify providers of the special billing and claims processing requirements for Zolgensma (onasemnogene abeparvovec-xioi) Suspension for intravenous infusion, when billed under a Healthcare Common Procedural Coding System (HCPCS) code, J3399. This communication supersedes the department's related communication, dated April 22, 2020.

Under the Healthcare Common Procedural Coding System (HCPCS), and effective July 1, 2020, Zolgensma was assigned the unique code, J3399 (Injection, onasemnogene abeparvovec-xioi, per treatment, up to 5×10^{15} vector genomes.). A non-specific HCPCS code, J3590, was used previously.

Coverage and policy details for Zolgensma under the Medi-Cal and California Children's Service (CCS) Programs are covered elsewhere.

National Standards and system limitations for J3399 do not allow for accurate claims adjudication when billing a single claim line. National Council for Prescription Drug Programs (NCPDP) standards and the UB-04 or other standard claim forms do not accommodate the large dollar amount of the claim, which is in excess of 2 million dollars.

When submitting claims for Zolgensma, providers are instructed to do the following:

1. Submit and receive back an approved Treatment Authorization Request (TAR) or approved product specific Service Authorization Request (SAR).
2. Bill using J3399, Injection, onasemnogene abeparvovec-xioi, per treatment, up to 5×10^{15} vector genomes.
3. Completion of Claim forms:
 - This billing methodology is restricted to hospital outpatient services. Note that pharmacies and clinics cannot bill using this methodology.
 - Outpatient claims may be billed electronically or by paper claim using 837I (Institutional) or UB-04 Medi-Cal claim forms with the following conditions:
 - The TAR/SAR is not negotiated.
 - Provider must submit one (1) service line on the TAR/SAR request, and enter "3" in the Units box.

- On the 837P or UB-04 claim form, provider must submit three (3) claim lines to represent one (1) service.
 - Each claim line to represent one unit.
 - Claims submitted with one or two claim lines will be denied.
 - Provider must submit an invoice for reimbursement.
 - This process will ensure that the total reimbursement paid for the three (3) claim lines is no more than the paid price on the provider submitted invoice.
 - Zolgensma must be billed on its own with no other drug or biological.
4. Providers are advised to take the following steps in order to ensure that Zolgensma claims are identified and processed expeditiously:

Paper claims may be identified by notation of “ Zolgensma “ on the “Remarks” section of the UB-04 claim form (Field #80) and submitted to:

Attention: Claims Manager
Medi-Cal Fiscal Intermediary/DXC
P.O. Box 526006
Sacramento, CA 95852-6006

Electronic claims may be identified by notation of ‘Zolgensma’ on the cover sheet, addressed to Attention: Claims Manager and submitted with the 837I claim form.

5. Providers to note that except for the first claim line, payment for any additional line will be delayed for 2-3 additional weeks due to systems constraints.
6. Payment for Zolgensma shall be a once in a lifetime reimbursement under J3399 or any other code (HCPCS, CPT, or by NDC).
7. For instructions regarding physician claim form completion, refer to the Medi-Cal website, forms section for completion of 837I form⁷ and UB-04 form⁸.

Below is a Zolgensma billing example using UB-04 form and with 3 claim lines:

- ✓ In this example, the total invoice cost of J3399 is \$2,125,002.00
- ✓ Note that each provider’s invoice cost may be different
- ✓ If this is split evenly between the 3 lines, each claim line will have a total of \$708,334.00
- ✓ The sum of the three claim lines must equal the paid price on the invoice
- ✓ Note that it is not necessary to include the unit of measure qualifier and numeric quantity.

Notice to Providers Regarding Special Billing

38		a b c			39 CODE	VALUE CODES AMOUNT	VALUE CODES AMOUNT	4 1 CODE	VALUE COD AMO
		d							
42 REV. CD.	43 DESCRIPTION	44 HCPCS / RATE / HIPPS CODE	45 SERV. DATE	46 SERV. UNITS	47 TOTAL CHARGES		48 NON-COVERED CHARGES	49	
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	N4 11digitNDC	J3399	070120	1	708,334	00			
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0001	PAGE 1 OF 1	CREATION DATE	070220	TOTALS	2,125,002	.00			
50 PAYER NAME	51 HEALTH PLAN ID	52 REL IN F O	53 ASG BEN	54 PRIOR PAYMENTS	55 EST. AMOUNT DUE	56 NPI	XXXXXXXXXX		

7 8371 Form
<https://mcweb.apps.prd.cammis.medi-cal.ca.gov/references/forms>
8 UB-04 Form
http://files.medi-cal.ca.gov/pubdoco/publications/masters-mtp/fpact/claimub_f00.doc