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

Coding and Billing Guide

Please see Indication and Important Safety Information, and accompanying Full Prescribing Information including Boxed Warning for Acute Serious Liver Injury and Acute Liver Failure.

An overview of this guide

This guide has been developed to assist you in obtaining insurance reimbursement for ZOLGENSMA® (onasemnogene abeparvovec-xioi) where health plans provide coverage for ZOLGENSMA as part of the medical benefit. As a result, 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	**LIMITED NETWORK SPECIALTY PHARMACIES**
Ordered from **CuraScript SD®** Phone: 866-263-8464	Novartis Gene Therapies has partnered with select Specialty Pharmacies to support ZOLGENSMA. Please contact your Novartis Gene Therapies Regional Account Associate Director (RAAD) or the OneGene Program® should you have any questions

This guide provides codes* for

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

*Codes provided in this guide may be subject to change. Please be sure to confirm the accuracy of all codes when submitting a claim.

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

*For live support about ordering, access, and reimbursement, or coding and billing, please contact your **Novartis Gene Therapies Regional Account Associate Director (RAAD)** with any questions you have.*

Indication and Important Safety Information

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

Indication

ZOLGENSMA® (onasemnogene abeparvovec-xioi) is an adeno-associated virus 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 (e.g., complete paralysis of limbs, permanent ventilator dependence) has not been evaluated with ZOLGENSMA.

Important Safety Information

BOXED WARNING: Acute Serious Liver Injury and Acute Liver Failure

Acute serious liver injury, acute liver failure, and elevated aminotransferases can 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 (e.g., hepatic aminotransferases [aspartate aminotransferase (AST) and alanine aminotransferase (ALT)], total bilirubin, and prothrombin time). Administer a systemic corticosteroid to all patients before and after ZOLGENSMA infusion. Continue to monitor liver function for at least 3 months after infusion.

WARNINGS AND PRECAUTIONS

Thrombocytopenia

Transient decreases in platelet counts, some of which met the criteria for thrombocytopenia, were typically observed within the first two 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 approximately 1 week after ZOLGENSMA infusion. Obtain baseline creatinine and complete blood count before ZOLGENSMA infusion. Following infusion, monitor for thrombocytopenia as well as other signs and symptoms of TMA. Consult a pediatric hematologist and/or pediatric nephrologist immediately to manage if 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.

ADVERSE REACTIONS

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

Please see accompanying Full Prescribing Information.

Dosing and NDC designation

The recommended dose of ZOLGENSMA® (onasemnogene abeparvovec-xioi) for single-dose intravenous 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 9 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 following tables list 22 ZOLGENSMA kits appropriate for dosing patients weighing between 2.6 kg and 13.5 kg.[1]

ZOLGENSMA Kit Sizes 2.6 kg to 13.5 kg

PATIENT WEIGHT (kg)	DOSE VOLUME[a] (mL)	KIT CONFIGURATION			NDC NUMBER
		5.5 mL VIALS	8.3 mL VIALS	# 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[b]	74.3	0	9	9	71894-141-09

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 13.5 kg.

[b] Dose volume for pediatric patients less than 2 years of age weighing equal to or greater than 13.6 kg will require a combination of ZOLGENSMA kits.

Some payers may require an 11 digit NDC code. In such cases, add a 0 in front of the second set of numbers, eg, 71894-141-09 would become 71894-**0141**-09.

Please see the ZOLGENSMA Treatment Guide for further dosing information.

Please see Indication and Important Safety Information, and accompanying Full Prescribing Information including Boxed Warning for Acute Serious Liver Injury and Acute Liver Failure.

HCPCS code

ZOLGENSMA® (onasemnogene abeparvovec-xioi) is billed under a HCPCS J-code. Effective July 1, 2020, the Centers for Medicare & Medicaid Services assigned ZOLGENSMA the following permanent J-code: J3399. HCPCS J-codes are used for items and supplies and non-physician services not covered by Current Procedural Terminology-4 (CPT-4) codes. J-codes are recommended when filling out and submitting a CMS-1450/UB-04 form.[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.

Utilization of the J-code is required when billing for ZOLGENSMA. Please ensure the billing system at your organization has been updated accordingly to support accurate coding and billing processes. The permanent J-code for ZOLGENSMA may also be found on the CMS website at: https://www.cms.gov/Medicare/Coding/HCPCSReleaseCodeSets/HCPCS-Quarterly-Update.

CPT and hospital revenue codes

ZOLGENSMA® (onasemnogene abeparvovec-xioi) is administered via a single-dose IV infusion.[1] The following Current Procedural Terminology (CPT) codes may be useful when coding and billing for ZOLGENSMA infusion. **Please note that these codes do not include office visits for diagnosis and prescribing of medication.** Appropriate codes can vary by setting of care, patient, and payer. It is the provider's responsibility to determine the appropriate health care setting and to submit true and correct claims for actual products and services rendered. Please check with the payer to verify codes and special billing requirements.

Remember, billing for ZOLGENSMA should be done separately and only if your institution is Buying and Billing. For facilities that choose to Buy and Bill, please refer to the section on HCPCS codes in this guide.

CPT codes for ZOLGENSMA infusion[4]

DESCRIPTION	CODE
IV infusion, for therapy, prophylaxis, or diagnosis; initial, up to one hour	96365
Initial observation care, per day, for the evaluation and management of a patient, which requires these 3 key components: a detailed or comprehensive history, a detailed or comprehensive examination, and medical decision-making that is straightforward or of low complexity, moderate complexity, or high complexity	99218-99220
Observation care admission and discharge services for the evaluation and management of a patient including admission and discharge on the same date, which requires these 3 key components: detailed or comprehensive history, a detailed or comprehensive examination, and medical decision-making that is straightforward or of low complexity, moderate complexity, or high complexity	99234-99236

Hospital revenue codes[5]

DESCRIPTION	CODE
IV therapy	026X
Incremental nursing care charges	0230 (general) 0231 (nursery)

You may need these codes regardless of the distribution method for ZOLGENSMA.

ICD-10 codes

A diagnosis of spinal muscular atrophy (SMA) is required by health plans to substantiate the medical necessity of ZOLGENSMA® (onasemnogene abeparvovec-xioi). Please refer to the following codes when documenting a patient's diagnosis.[6]

ICD-10 codes for SMA[6]

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 see Potential Health Plan Prior Authorization Criteria in the Reimbursement Resources binder for more information about anticipated health plan requirements.

CPT codes for testing

Insurance providers require the following tests prior to infusion with ZOLGENSMA® (onasemnogene abeparvovec-xioi):

- SMA Diagnostic Test which includes: *SMN1* deletion and *SMN2* copy number
- AAV9 Antibody Test

Monitoring is required prior to and post-infusion with ZOLGENSMA

Perform baseline tests for liver function, creatinine, complete blood count (including hemoglobin and platelet count), and troponin-I. Post-infusion monitor liver function, platelet count and troponin-I on a regular basis for at least 3 months, as described in the Prescribing Information.

Novartis Gene Therapies covers the cost for the following tests as part of the Novartis Gene Therapies Laboratory Testing Program:

- SMA Diagnostic Test which includes: *SMN1* deletion and *SMN2* copy number
- AAV9 Antibody Test

If you have any questions regarding the Novartis Gene Therapies Laboratory Testing Program, please contact the OneGene Program® at 855-441-GENE (4363), Monday-Friday (8 AM to 8 PM ET), for additional assistance.

Additional recommended testing codes related to treatment with ZOLGENSMA[4]

TEST	CODE*
AST value test	84450
ALT value test	84460
Troponin-I test	84484
CBC test (includes differential and platelets)	85025
Bilirubin, total	82247
Prothrombin time	85610
Creatinine	82565

*A specific test code may be required in addition to the CPT code. Please confirm which codes are required for your preferred laboratory.

*Your **Novartis Gene Therapies Regional Account Associate Director (RAAD)** can answer any questions you may have about lab tests. You may also call Novartis Gene Therapies Medical Information at 833-828-3947.*

Submitting forms and claims

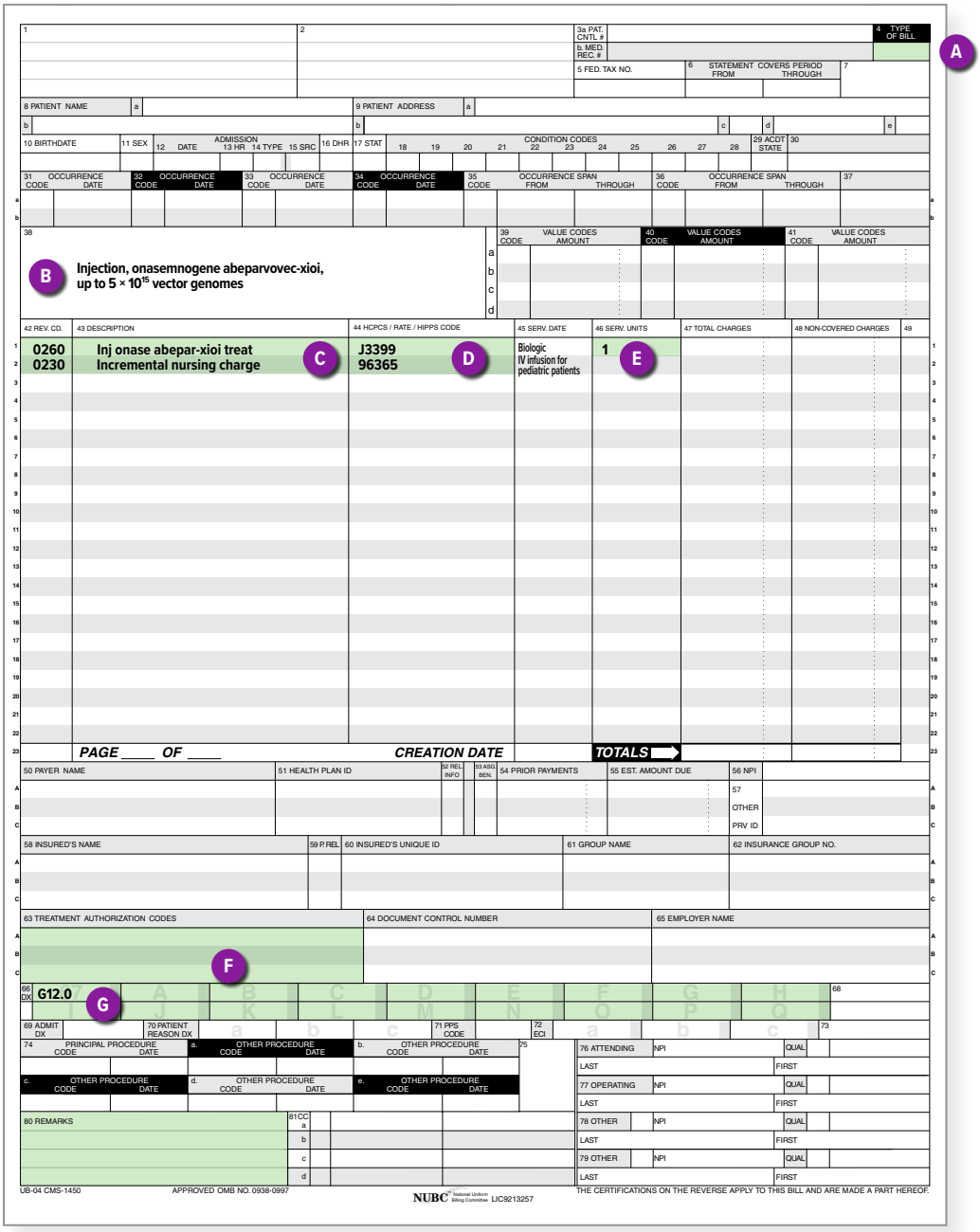
When submitting claims for ZOLGENSMA® (onasemnogene abeparvovec-xioi), you will most likely be required to fill out the CMS 1450/UB-04 Form. The following suggestions can help with the claim process:

- ☑ Ensure the following information is accurate:
  - Patient name and date of birth
  - Patient's parent/legal guardian name, ID number, address
    - Please note that in the first 30 days of life, the patient may be listed under the mother's name until the baby is independently insured
  - Prescriber name and National Provider Identifier (NPI)
  - ICD-10 code
- ☑ Confirm that your patient's medical records support the diagnosis and procedure codes
- ☑ Include a brief description of the medical reason for treatment and why ZOLGENSMA is medically necessary
- ☑ Enter the prior authorization number provided by your patient's insurance carrier

*Please contact your **Novartis Gene Therapies Regional Account Associate Director (RAAD)** with any questions you have about ZOLGENSMA coding and billing.*

Sample CMS-1450/UB-04 form

Please consider using the following fields and codes when billing for outpatient hospital services:



A **Box 4:** Enter the appropriate type of bill code; for example
- 013X, Hospital outpatient
- 014X, Hospital other Part B

B **Box 38:** Add the description for the J-code 3399: Injection, onasemnogene abeparvovec-xioi, up to 5 × 10^{15} vector genomes

C **Boxes 42 and 43:** Enter appropriate revenue code description of service

D **Box 44:** Enter J3399

E **Box 46:** Enter the number of units administered

F **Box 63:** Indicate the prior authorization number here

G **Box 67:** Enter the appropriate ICD-10 diagnosis code that corresponds to the patient's diagnosis, such as G12.0 (Infantile SMA, Type I [Werdnig-Hoffman])

Appropriate codes can vary by setting of care, patient, and payer. It is the provider's responsibility to determine the appropriate health care setting and to submit true and correct claims for actual products and services rendered. Please check with the payer to verify codes and special billing requirements.

Please see Indication and Important Safety Information, and accompanying Full Prescribing Information including Boxed Warning for Acute Serious Liver Injury and Acute Liver Failure.

Frequently asked questions

How to Order

1. How do I order ZOLGENSMA® (onasemnogene abeparvovec-xioi)?

For assistance with ordering and billing, please contact your Novartis Gene Therapies Regional Account Associate Director (RAAD) who will be able to provide you with materials and information to address the specific needs of your office.

ZOLGENSMA may be ordered in 2 ways:

- **Purchase directly from CuraScript SD®.** Order the appropriate ZOLGENSMA kit, and CuraScript SD® will drop-ship directly to your treatment center pharmacy
- **Order through one of the limited network Specialty Pharmacies.** The Specialty Pharmacy will order the patient-specific ZOLGENSMA kit, bill your patient's insurance, and ship product to your treatment center pharmacy

Insurance Coverage

2. Is ZOLGENSMA covered under an inpatient diagnosis-related group (DRG) for billing?

- ZOLGENSMA is intended to be administered in an outpatient setting and is therefore not expected to be reimbursed through DRG payment structures
- Please contact the patient's health plan as needed for additional clarity

3. Does Novartis Gene Therapies cover the cost of the diagnostic testing?

The Novartis Gene Therapies Laboratory Testing Program covers the cost of the following diagnostic lab tests:

- SMA Diagnostic Test which includes: *SMN1* deletion and *SMN2* copy number
- AAV9 Antibody Test

To learn more, please contact your Novartis Gene Therapies Regional Account Associate Director (RAAD)

4. How do I bill for ZOLGENSMA through state Medicaid?

Each Medicaid plan will vary in their coverage policies. Please contact your Novartis Gene Therapies Regional Account Associate Director (RAAD) for a detailed discussion of state policies.

5. How does insurance reimburse for infusion of ZOLGENSMA?

For Buy and Bill treatment centers, the cost of ZOLGENSMA should be billed separately from services related to its infusion. Please refer to the CPT and hospital revenue codes in this guide for more information that will help you when billing for infusion of ZOLGENSMA.

Frequently asked questions

Infusion

6. Where should ZOLGENSMA® (onasemnogene abeparvovec-xioi) be infused?

Please call the OneGene Program® at 855-441-GENE (4363) for more information.

7. How long is the ZOLGENSMA infusion?

ZOLGENSMA is administered as a slow infusion over approximately 60 minutes. Do not infuse as an IV push or bolus. Once the dose is drawn into the syringe, it must be used within 8 hours.[1]

8. How do I prepare the patient for infusion?

ZOLGENSMA should only be infused by a health care provider. Prior to ZOLGENSMA infusion, health care providers should assess liver function, measure creatinine, complete blood count (including hemoglobin and platelet count), and troponin-I, and perform baseline testing for the presence of anti-AAV9 antibodies.

Postpone ZOLGENSMA in patients with concurrent infections until the infection has resolved. Clinical signs or symptoms of infection should not be evident at the time of ZOLGENSMA administration.

To manage a possible increase in liver transaminases, all patients should receive prednisolone given orally.*[1] If oral corticosteroid therapy is not tolerated, consider intravenous corticosteroids as clinically indicated.

- Order prednisolone for the patient and discuss infusion with the family. One day prior to infusion, begin administration of oral prednisolone at a dose of approximately 1 mg/kg/day for a total of 30 days
- Monitor liver function by clinical examination and by laboratory testing, including assessing ALT, AST, total bilirubin, and prothrombin time, on a regular basis for at least 3 months (weekly for the first month, and every other week for the second and third month until results are unremarkable)
  - Following 30 days of prednisolone treatment, the 1 mg/kg/day dose can be tapered gradually over 28 days for patients with unremarkable findings (normal clinical exam, total bilirubin, and prothrombin time, and ALT and AST levels below 2 x upper limit of normal [(ULN]). Do not stop systemic corticosteroids abruptly.
  - If abnormalities persist, continue prednisolone treatment until findings become unremarkable, and then taper the corticosteroid dose gradually over the next 28 days or longer if needed. Do not stop systemic corticosteroids abruptly. Consult a pediatric gastroenterologist or hepatologist.
  - Platelet counts weekly for the first month, and then every other week for the second and third months, until platelet counts return to baseline
  - Troponin-I weekly for the first month, and then monthly for the second and third months, until troponin-I level returns to baseline

9. Is the prednisolone pretreatment mandatory or optional? What if my patient is allergic to prednisolone?

Prednisolone pretreatment is recommended and may be required by the patient's health plan. Consult expert(s) if patients do not respond adequately to the equivalent of 1 mg/kg/day oral prednisolone.

Acute serious liver injury and acute liver failure can occur with ZOLGENSMA. Prior to ZOLGENSMA infusion, health care providers should assess liver function, measure creatinine, complete blood count (including hemoglobin and platelet count), and troponin-I, and perform baseline testing for the presence of anti-AAV9 antibodies. Liver function should be monitored by clinical examination and by laboratory testing for at least 3 months.

Monitor liver function by clinical examination and by laboratory testing on a regular basis, including assessing ALT, AST, total bilirubin, and prothrombin time. Variance from these recommendations is at the discretion of the treating physician.

*Systemic corticosteroids equivalent to oral prednisolone at 1 mg/kg/day can be used.

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*For live support about ordering, access, and reimbursement, or coding and billing, please contact your **Novartis Gene Therapies Regional Account Associate Director (RAAD)** with any questions you have.*

References: 1. ZOLGENSMA [prescribing information]. Bannockburn, IL: Novartis Gene Therapies, Inc.; 2021. **2.** HCPCS release & code sets. Centers for Medicare & Medicaid Services. https://www.cms.gov/Medicare/Coding/HCPCSReleaseCodeSets. Accessed November 1, 2021. **3.** HCPCS quarterly update. Centers for Medicare & Medicaid Services website. https://www.cms.gov/Medicare/Coding/HCPCSReleaseCodeSets/HCPCS-Quarterly-Update. Accessed November 1, 2021. **4.** American Medical Association. CPT® Professional 2020 Edition. Chicago, IL: American Medical Association; 2020. **5.** Centers for Medicare & Medicaid Services. CMS Manual Update: Pub. 100-04 Medicare Claims Processing. https://www.cms.gov/Regulations-and-Guidance/Guidance/Transmittals/Downloads/R167CP.pdf. Accessed November 1, 2021. **6.** ICD-10-CM tabular list of diseases and injuries. Centers for Medicare & Medicaid Services website. https://www.cms.gov/medicare/icd-10/2021-icd-10-pcs. Accessed November 1, 2021.

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