There for Rare
by Immedica Pharma

CODING & BILLING GUIDE

for Loargys® (pegzilarginase-nbln)

A reference guide for diagnostic, administration, product, and revenue codes that can help payers recognize, process, and pay claims for LOARGYS.

IMPORTANT SAFETY INFORMATION

WARNING: HYPERSENSITIVITY REACTIONS INCLUDING ANAPHYLAXIS

See full prescribing information for complete boxed warning

Initiate LOARGYS in a healthcare setting with appropriate medical monitoring and support measures, including access to cardiopulmonary resuscitation equipment. If a severe hypersensitivity reaction (e.g., anaphylaxis) occurs, discontinue LOARGYS, and immediately initiate appropriate medical treatment, including use of epinephrine.

WARNINGS AND PRECAUTIONS

Hypersensitivity Reactions Including Anaphylaxis: Life-threatening hypersensitivity reactions, including anaphylaxis, have occurred in patients treated with enzyme replacement therapies, including LOARGYS. Hypersensitivity reactions that were mild to moderate in severity occurred in 13% (6/48) of LOARGYS-treated subjects in clinical trials. Hypersensitivity reactions have included facial swelling, rash, flushing and dyspnea. The reactions generally occurred with the first few doses but may occur later in treatment.

Please see additional Important Safety Information on following pages and full Prescribing Information.

Loargys®
(pegzilarginase-nbln)

A guide to coding, billing, and reimbursement for Loargys® (pegzilarginase-nbln)

This resource provides **coding and billing information** to help with the **reimbursement process for LOARGYS administered in the office or hospital outpatient setting.**

This guide includes:

- **Diagnostic coding:** International Classification of Diseases, 10th Revision, Clinical Modification (ICD-10-CM) codes
- **Administration coding:** Current Procedural Terminology (CPT) codes*
- **Product coding:** Healthcare Common Procedure Coding System (HCPCS) Level II codes
- **Revenue codes**

The coding information discussed in this guide is provided for informational purposes only, is subject to change and interpretation, and should not be construed as legal advice. The codes listed herein may not apply to all patients or to all health plans. Conversely, additional codes not listed in this guide may apply to some patients. Providers should follow payer-specific coding requirements and exercise independent clinical judgement when selecting codes and submitting claims to accurately reflect the services and products furnished to a specific patient. Providers must determine whether it is appropriate to submit any particular claim for reimbursement. Information provided in this guide is effective as of October 2026.

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IMPORTANT SAFETY INFORMATION (CONTINUED)
WARNINGS AND PRECAUTIONS (CONTINUED)

Administration of LOARGYS should be supervised by a healthcare provider knowledgeable in the management of hypersensitivity reactions including anaphylaxis in a healthcare setting with appropriate medical monitoring and support measures. Premedication with an antihistamine and/or corticosteroid should be considered in patients who previously have developed a hypersensitivity reaction. If a *severe* hypersensitivity reaction (e.g., anaphylaxis) occurs, discontinue LOARGYS and immediately initiate appropriate medical treatment, including use of epinephrine. Consider the risks and benefits of re-administering LOARGYS in patients who have experienced a severe hypersensitivity reaction. Caution should be exercised upon rechallenge. Inform patients of the symptoms of life-threatening hypersensitivity reactions and to seek immediate medical attention should symptoms occur. If a mild or moderate reaction occurs, consider treatment with antihistamines and/or corticosteroids.

Please see additional Important Safety Information on following pages and full Prescribing Information.

Commonly used billing codes

This table shows common billing codes that may be used when filing claims for Loargys® (pegzilarginase-nbln). These codes are provided for informational purposes only; please contact your individual payers to determine appropriate codes and billing requirements.

ICD-10-CM	**E72.21**	Argininemia

CPT codes	**96365**	Intravenous infusion services for therapy prophylaxis or diagnosis purposes up to 1 hour
	96372	Therapeutic, prophylactic, or diagnostic injection (specify substance or drug); subcutaneous or intramuscular
	G0463	Hospital outpatient clinic visit for assessment and management of a patient
	G03778	Hospital observation service, per hour
	G0379	Direct admission of patient for hospital observation care

LOARGYS, when administered in the hospital outpatient setting, may require observational services. Observation requires a series of clinically appropriate services like short-term treatment, assessment, and reassessment of the patient. Claims reporting observation services must also include an initial service CPT such as a clinic or emergency department visit. Medical observation records should contain dated and time-stamped physician's orders describing the observation services the patient is to receive. Refer to specific payer requirements before reporting observation codes.

J codes	J1818	Injection, pegzilarginase-nbln, 0.05 mg

Effective July 1, 2023, CMS and most payers require prescribers to record drug wastage, or lack thereof. A JW modifier may be required for reporting that there was discarded drug (JXXXX-JW). A JZ modifier may be required for reporting there was no discarded drug (JXXXX-JZ).

Revenue codes allow hospitals to capture cost data for billing of services provided.

Revenue code	Description
Administration	
0510	Clinic
0500	Outpatient services
Drug	
0636	Drugs requiring detailed coding
0250	Drugs and biologicals
0260	IV therapy

Drug name/strength	10-digit NDC #	11-digit NDC #*
LOARGYS 2 mg/0.4 mL	81583-102-01	81583-**0**102-01

*The product's NDC has been 'zero-filled' to ensure creation of an 11-digit code that meets general billing standards. The zero-fill is indicated in bold.

IMPORTANT SAFETY INFORMATION (CONTINUED)
ADVERSE REACTIONS

The most common adverse reactions are vomiting, pyrexia, infusion associated reactions and constipation.

Please see additional Important Safety Information on following pages and full Prescribing Information.

Common billing claim forms

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Two of the most common billing claim forms are listed below and are available from the Centers for Medicare & Medicaid Services (CMS) at https://www.cms.gov/medicare/forms-notices/cms-forms-list:

- **CMS-1500** (print) or **837P** (electronic) for physician office reimbursement

- **UB-04** (also known as CMS-1450) (print) or **8371** (electronic) for hospital outpatient reimbursement

Sample annotated versions of these forms can be found on the following pages

IMPORTANT SAFETY INFORMATION (CONTINUED)
USE IN SPECIFIC POPULATIONS

Pregnancy: There are no available data on LOARGYS use in pregnant females to evaluate for a drug-associated risk of major birth defects, miscarriage or other adverse maternal or fetal outcomes.

Please see additional Important Safety Information on following pages and full Prescribing Information.

Loargys® (pegzilarginase-nbln)

Sample annotated CMS-1500 form*

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The CMS-1500 form is commonly used for billing for Loargys® (pegzilarginase-nbln) when it is administered in physician offices.

This sample claim form is intended for informational purposes only. The coding information provided is subject to change and should not be construed as billing advice. Use of the coding information provided is not a guarantee that reimbursement will be provided. Providers are responsible for ensuring that claims submitted are appropriate and accurately reflect the services and products furnished to a specific patient. Please confirm the accuracy of the codes you use to bill for LOARGYS with each payer.

Box 19 Some payers require additional information in Box 19. Additional information may include method of administration, the name of the drug, and the total dose. Verify specific requirements with your payer. If you cannot provide an adequate description within this box, provide an attachment to your claim

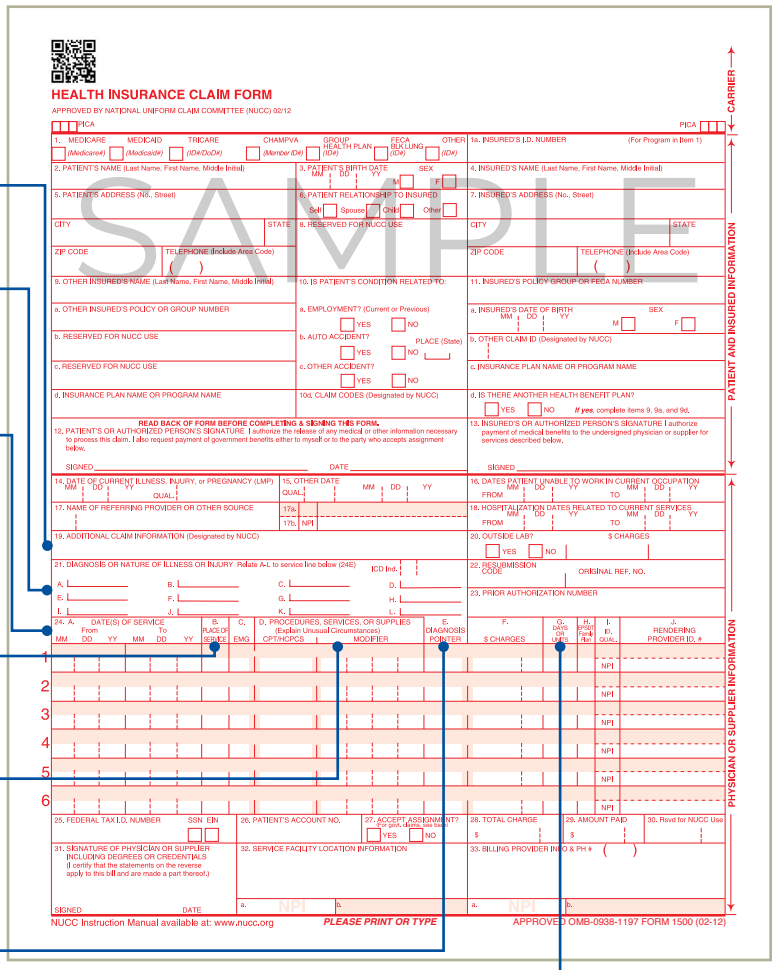
Box 21 Enter appropriate ICD-10-CM codes for the patient's diagnosis

Box 24A Enter a 6- or 8-digit (month-day-year) date for each procedure, service, or supply. In the red shaded area, an NDC is also required for LOARGYS. Submit "N4" followed by the 11-digit NDC, quantity qualifier, and units of measure

Box 24B Enter the appropriate place of service code

Box 24D Enter the appropriate CPT and HCPCS codes and modifiers for procedures, services, and supplies, as required by the specific payer. Enter HCPCS code J1818. In addition, Medicare and most payers require you to record drug waste. Enter HCPCS code J1818 and use modifier JZ if zero drug was wasted (ie, J1818-JZ) or record waste on a separate line with the JW modifier (ie, J1818-JW)

Box 24E Enter the diagnosis code reference letter (as appropriate, per form version) as shown in item 21 to relate the date of service and the procedures performed to the primary diagnosis. Enter only 1 reference letter per line item. If multiple services were performed, enter the primary reference letter for each service



Box 24G Enter the number of LOARGYS units administered. If a separate line item was created for waste as J1818-JW, clearly indicate the number of units discarded. Both lines will be processed for payment

CMS, Centers for Medicare & Medicaid Services; CPT, Current Procedural Terminology; HCPCS, Healthcare Common Procedure Coding System; ICD-10-CM, International Classification of Diseases, Tenth Revision, Clinical Modification; NDC, National Drug Code.

*CMS 837P (not shown) is the electronic equivalent of CMS-1500. It should be used if you submit your claims electronically.

IMPORTANT SAFETY INFORMATION (CONTINUED)
USE IN SPECIFIC POPULATIONS (CONTINUED)

Lactation: There is no data on the presence of LOARGYS in either human or animal milk, the effects on the breastfed infant, or the effects on milk production. The developmental and health benefits of breastfeeding should be considered along with the mother's clinical need for LOARGYS and any potential adverse effects on the breast-fed infant from LOARGYS or from the underlying maternal condition.

Please see additional Important Safety Information on following pages and full Prescribing Information.

Loargys® (pegzilarginase-nbln)

Example scenarios for Loargys® (pegzilarginase-nbln) using the CMS-1500 form

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Below are some examples of dosing scenarios for LOARGYS and how they may be recorded on the CMS-1500 form. Check with the applicable payer for specific requirements.

1. LOARGYS is supplied in 2 mg/0.4 mL single-dose vials (1 single-dose vial=40 billing units)
2. The recommended starting dosage of LOARGYS is 0.1 mg/kg (actual body weight) administered via intravenous infusion once weekly
3. Store refrigerated at 2 °C to 8 °C (36 °F to 46 °F) in the original carton to protect from light. Do not freeze. Do not shake

Please see full Prescribing Information for more directions on proper dosing and administration.

Dosing Example 1

Patient Weight: 11.33 kg (25 lb)
LOARGYS Dose: 0.1 mg/kg
Total Dose Required: 1.133 mg (0.2 mL)
LOARGYS 2 mg/0.4 mL single-dose vials required: 1

- **Box 24 A:** In the red shaded area, the NDC should be submitted using "N4" followed by the 11 digits, quantity qualifier, and units of measure; in this case, N4815830102011ML0.2
- **Box 24 G:** A 1.133 mg dose would be billed as "J1818: 22.66 units; J1818-JW: 17.34 units" (0.05 mg of LOARGYS=1 billing unit)

Dosing Example 2 (Titration)

Patient Weight: 22.68 kg (50 lb)
LOARGYS Dose: 0.15 mg/kg
Total Dose Required: 3.402 mg (0.7 mL)
LOARGYS 2 mg/0.4 mL single-dose vials required: 2

- **Box 24 A:** In the red shaded area, the NDC should be submitted using "N4" followed by the 11 digits, quantity qualifier, and units of measure; in this case, N4815830102011ML0.7
- **Box 24 G:** A 3.402 mg dose would be billed as "J1818: 68.04 units; J1818-JW: 11.96 units" (0.05 mg of LOARGYS=1 billing unit)

Dosing Example 3 (Titration)

Patient Weight: 68.04 kg (150 lbs)
LOARGYS Dose: 0.2 mg/kg
Total Dose Required: 13.608 mg (2.7 mL)
LOARGYS 2 mg/0.4 mL single-dose vials required: 7

- **Box 24 A:** In the red shaded area, the NDC should be submitted using "N4" followed by the 11 digits, quantity qualifier, and units of measure; in this case, N4815830102011ML2.7
- **Box 24 G:** A 13.608 mg dose would be billed as "J1818: 272.16 units, J1818-JW: 7.84 units" (0.05 mg of LOARGYS=1 billing unit)

"White-Bagging" Scenario

When a LOARGYS prescription is filled by a third party, such as a specialty pharmacy, the provider only bills for the administration of the product. The product's J-code should still be included on the form, but the rate should be recorded as "0" or a nominal amount (eg, $0.01), or as accepted by the payer

Patient Weight: 58.97 kg (130 lb)
LOARGYS Dose: 0.1 mg/kg
Total Dose Required: 5.897 mg (1.2 mL)
LOARGYS 2 mg/0.4 mL single-dose vials required: 3

- **Box 24 A:** In the red shaded area, the NDC should be submitted using "N4" followed by the 11 digits, quantity qualifier, and units of measure; in this case, N4815830102011ML1.2
- **Box 24 F:** In a white-bagging scenario, enter 0 or a nominal amount for the charges
- **Box 24 G:** A 5.897 mg dose would be billed as "J1818: 117.94 units; J1818-JW: 2.06 units" (0.05 mg of LOARGYS=1 billing unit)

IMPORTANT SAFETY INFORMATION (CONTINUED)
USE IN SPECIFIC POPULATIONS (CONTINUED)

Pediatric: The safety and effectiveness of LOARGYS have been established for the reduction of plasma arginine in pediatric patients 2 years and older with ARG-1 D, in conjunction with dietary protein restriction. The safety and effectiveness of LOARGYS have not been established for the reduction of plasma arginine in pediatric patients aged less than 2 years with ARG-1 D.

Please see additional Important Safety Information on following pages and full Prescribing Information.

Loargys® (pegzilarginase-nbln)

Sample annotated CMS UB-04 form*

There for Rare by Immedica Pharma

The CMS UB-04 form is commonly used for billing for Loargys® (pegzilarginase-nbln) when it is administered in hospital outpatient settings.

This sample claim form is intended for informational purposes only. The coding information provided is subject to change and should not be construed as billing advice. Use of the coding information provided is not a guarantee that reimbursement will be provided. Providers are responsible for ensuring that claims submitted are appropriate and accurately reflect the services and products furnished to a specific patient. There are 81 fields or lines on a UB-04 form and they are referred to as Form Locators (FL). Please confirm the accuracy of the codes you use to bill for LOARGYS with each payer.

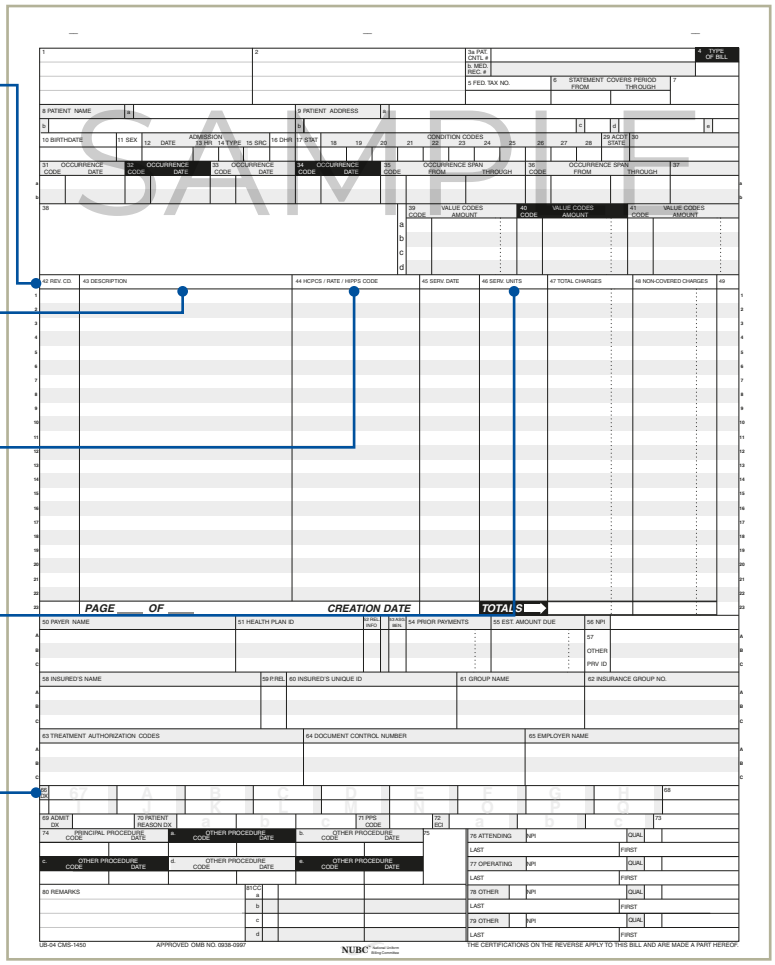
FL 42 List revenue codes in ascending order. Enter the appropriate numeric revenue code in FL 42 to explain each charge in FL 47

FL 43 For each revenue code reported in FL 42, provide a narrative description or standard abbreviation in FL 43. If an NDC is required, submit "N4" followed by the 11-digit NDC, quantity qualifier, and units of measure

FL 44 Enter the appropriate CPT and HCPCS codes and modifiers for procedures, services, and supplies as required by the specific payer. Medicare and most payers require drug waste to be recorded. Enter HCPCS code J1818 and use modifier JZ if zero drug was wasted (ie, J1818-JZ), or record waste on a separate line with the JW modifier (ie, J1818-JW)

FL 46 Enter the number of LOARGYS units administered. If a separate line item was created for waste as J1818-JW, clearly indicate the number of units discarded. Both lines will be processed for payment

FL 67 Enter the appropriate ICD-10-CM diagnosis code(s) for the patient's diagnosis



*CMS 837I (not shown) is the electronic equivalent of CMS-1450/UB-04. It should be used if you submit your claims electronically.

IMPORTANT SAFETY INFORMATION (CONTINUED)
USE IN SPECIFIC POPULATIONS (CONTINUED)

Geriatric: Clinical studies of LOARGYS did not include subjects 65 years of age and older to determine whether they respond differently from younger adult subjects.

Loargys® (pegzilarginase-nbln)

Please see additional Important Safety Information on following pages and full Prescribing Information.

Example scenarios for Loargys® (pegzilarginase-nbln) using the CMS UB-04 form

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Please see the below example dosing scenarios for LOARGYS and how they may be recorded on the CMS UB-04 form. Check with the applicable payer for specific requirements.

1. LOARGYS is supplied in 2 mg/0.4 mL single-dose vials (1 single-dose vial=40 billing units)
2. The recommended starting dosage of LOARGYS is 0.1 mg/kg (actual body weight) administered via intravenous infusion once weekly
3. Store refrigerated at 2 °C to 8 °C (36 °F to 46 °F) in the original carton to protect from light. Do not freeze. Do not shake

Please see full Prescribing Information for more directions on proper dosing and administration.

Dosing Example 1

Patient Weight: 36.29 kg (80 lb)

LOARGYS Dose: 0.1 mg/kg

Total Dose Required: 3.629 mg (0.7 mL)

LOARGYS 2 mg/0.4 mL single-dose vials required: 2

- **FL 42 and 43:** Insert the revenue codes in ascending order. In this case, 0500 was used for the Outpatient service and 0636 was used for the product. The NDC should be submitted using "N4" followed by 11 digits, quantity qualifier, and units of measure; in this case, N4815830102011ML0.7
- **FL 45:** Include date of service
- **FL 46:** A 3.629 mg dose would be billed as "J1818: 72.58 units; J1818-JW: 7.42 units" (0.05 mg of LOARGYS=1 billing unit)

Dosing Example 2 (Titration)

Patient Weight: 68.04 kg (150 lb)

LOARGYS Dose: 0.2 mg/kg

Total Dose Required: 13.608 mg (2.7 mL)

LOARGYS 2 mg/0.4 mL single-dose vials required: 7

- **FL 42 and 43:** Insert the revenue codes in ascending order. In this case, 0510 was used for the clinic and 0636 was used for the product. The NDC should be submitted using "N4" followed by 11 digits, quantity qualifier, and units of measure; in this case, N4815830102011ML2.7
- **FL 45:** Include date of service
- **FL 46:** A 13.608 mg dose would be billed as "J1818: 272.16 units; J1818-JW: 7.84 units" (0.05 mg of LOARGYS=1 billing unit)

IMPORTANT SAFETY INFORMATION (CONTINUED)
INDICATION

LOARGYS is an arginine specific enzyme indicated for the treatment of hyperargininemia in adult and pediatric patients 2 years of age and older with Arginase 1 Deficiency (ARG1-D), in conjunction with dietary protein restriction.

This indication is approved under accelerated approval based on reduction of plasma arginine. Continued approval for this indication may be contingent upon verification and description of clinical benefit in a confirmatory trial.

Please see additional Important Safety Information on previous pages and full Prescribing Information.

Loargys® (pegzilarginase-nbln)

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If you have questions about coding and billing for Loargys® (pegzilarginase-nbln), please call us at 1-844-982-5691 and select Option 2, Monday–Friday, 8AM–8PM Eastern time

References: 1. Centers for Medicare & Medicaid Services. Medicare claims processing manual. Chapter 26: completing and processing form CMS-1500 data set. Updated August 9, 2024. Accessed July 10, 2025. https://www.cms.gov/Regulations-and-Guidance/Guidance/Manuals/Downloads/clm104c26.pdf **2.** Centers for Medicare & Medicaid Services. Medicare claims processing manual. Chapter 25: completing and processing the form CMS-1450 data set. Updated December 20, 2023. Accessed July 10, 2025. https://www.cms.gov/Regulations-and-Guidance/Guidance/Manuals/Downloads/clm104c25.pdf

Please see additional Important Safety Information on previous pages and full Prescribing Information.

Immedica pharma

Loargys® (pegzilarginase-nbln)

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