Lembaz™
(adrabetadex) injection
900 mg/4.5 mL, for intrathecal use

Relevant Codes and Sample Claim Forms for Lembaz

This brochure is designed to support your practice or site of care with accurate and efficient claim preparation for Lembaz in medical buy-and-bill settings. It provides an overview of best practices to assist with coding for procedures related to Lembaz administration and ancillary services (if needed), and examples of claim forms and guidance on how to complete them. This brochure is not meant to provide medical guidance. It is up to the provider to determine appropriate medical treatment and code accordingly.

INDICATION AND IMPORTANT SAFETY INFORMATION

INDICATION

LEMBAZ™ (adrabetadex) is indicated to reduce the risk of mortality in adult and pediatric patients, who weigh 7 kg or greater, with Niemann-Pick disease type C (NPC) with neurological manifestations that started at less than 6 years of age.

IMPORTANT SAFETY INFORMATION

WARNING: HEARING IMPAIRMENT INCLUDING PERMANENT HEARING LOSS

Treatment with LEMBAZ has resulted in permanent hearing impairment.

- **Prior to LEMBAZ use, perform a comprehensive audiology evaluation (e.g., pure tone audiometry, speech recognition, tympanometry) to establish baseline hearing levels.**
- **During LEMBAZ treatment, modify LEMBAZ dosage based on comprehensive audiology evaluations as follows:**
  - **To determine the subsequent dosage level, repeat comprehensive audiology evaluation 1 to 2 weeks after the first LEMBAZ dose at each dosage level but before the next dose, and then 1 to 2 weeks after the second LEMBAZ dose at each dosage level but before the next dose.**
  - **When a patient remains at the same LEMBAZ dosage level for 2 consecutive doses or has reached the maximum dosage, perform comprehensive audiology evaluations at one-month intervals until 3 consecutive, stable audiology results are obtained, and then repeat the comprehensive audiology evaluation every 12 months.**

Please see additional Important Safety Information on page 12 and the full Prescribing Information, including Boxed WARNING.

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Sample Coding for Lembaz™ (adrabetadex)

Remember, billing and coding recommendations may vary by health plan. Your practice or facility should check directly with the patient's health plan(s) for guidance on the appropriate codes to use to facilitate claim processing for Lembaz, its administration, and any ancillary services.

ICD-10-CM code examples[1]

ICD-10-CM codes are used to identify and document a patient's diagnosis, condition, symptom, or reason for receiving care.

ICD-10-CM code	Description
E75.24	Niemann-Pick disease
E75.242	Niemann-Pick disease, type C
E75.249	Niemann-Pick disease, unspecified

HCPCS code[2]

At this time, Lembaz does not have a product-specific HCPCS code. Until a product-specific HCPCS code is assigned and recognized by payers, HCPs should confirm the appropriate miscellaneous or unclassified drug/biologic code with the payer and the site's coding or revenue-cycle team. Candidate codes may differ by site of care.

When using a miscellaneous or unclassified code, include the drug name, dose, and NDC number on the claim when required. C9399, J3490, and J3590 are not interchangeable. Code selection depends on payer requirements, site of care, product classification, and HCP coding/revenue-cycle review. Payers may require additional documentation to process claims.

HCPCS code	Description
C9399*	Unclassified drugs or biologics. Hospital outpatient/OPPS-oriented use; payer and facility validation required.
J3490	Unclassified drugs. Professional/CMS-1500 or payer-directed miscellaneous drug pathway.
J3590	Unclassified biologics. Unclassified biologic pathway only if payer direction and product classification support it.

340B and drug wastage modifiers[3-5]

340B refers to the 340B Drug Pricing Program, which requires participating manufacturers to provide outpatient drugs to eligible healthcare organizations at a discounted price. A wastage modifier is used when an HCP must discard part of a single-use vial because the entire amount cannot be administered to the patient.

Modifiers should be appended only when supported by the service performed, site of care, payer requirements, and documentation. Drug wastage, 340B, E/M, anesthesia, reduced/discontinued service, multiple procedure, and distinct procedural service modifiers follow different rules and are not interchangeable.

Modifiers	Description
Drug wastage/ no wastage	JW and JZ apply to the drug line when required. JW indicates discarded amount while JZ indicates no discarded amount. They are not simultaneously applicable to the same drug-line logic.
JW	Amount discarded/not administered to any patient, when required for the drug line.
JZ	No discarded amount, when required for the drug line.
TB	340B reporting where applicable, including current Medicare reporting where required.
UD	Medicaid or payer-specific 340B reporting when required; not equivalent to TB.

E/M=evaluation and management; HCPCS=Healthcare Common Procedure Coding System; ICD-10-CM=*International Classification of Diseases, Tenth Revision, Clinical Modification*; OPPS=Outpatient Prospective Payment System; NDC=National Drug Code.

*This code is only to be used by facilities.

Please see additional Important Safety Information on page 12 and the full Prescribing Information, including Boxed WARNING.

Sample Coding for Lembaz™ (adrabetadex) (cont'd)

NDC number[6]

An NDC number is product-identifying information and is not a substitute for HCPCS, CPT, diagnosis, revenue, or ICD-10-PCS coding. When NDC reporting is required, the NDC format, unit of measure, and quantity should follow payer, Medicaid, and 837 companion-guide requirements.

NDC number		Description
10-digit format	**11-digit format**	**Note: Both formats are listed for reference. NDC reporting format, unit of measure, and quantity should follow payer, Medicaid, and 837 companion-guide requirements.**
87230-101-01	87230-0101-01	One single-dose vial

ICD-10-PCS procedure codes[7]

The ICD-10-PCS is a procedure classification system used for inpatient facility procedure reporting. ICD-10-PCS codes are not substitutes for CPT or HCPCS codes on outpatient or professional claims.

ICD-10-PCS	Description
009U3ZX	Drainage of cerebrospinal fluid, spinal canal, percutaneous approach, diagnostic
009U3ZZ	Drainage of cerebrospinal fluid, spinal canal, percutaneous approach
BR13YZZ	Fluoroscopy of lumbar disc(s) using other contrast
BR49ZZZ	Ultrasonography of lumbar spine

CPT=Current Procedural Terminology; HCPCS=Healthcare Common Procedure Coding System; ICD-10-PCS=*International Classification of Diseases, Tenth Revision, Procedure Coding System*; NDC=National Drug Code.

Please see additional Important Safety Information on page 12 and the full Prescribing Information, including Boxed WARNING.

Sample Coding for Lembaz™ (adrabetadex) (cont'd)

CPT® code examples[8,9]

All CPT codes should be validated by the HCP to ensure accurate coding. These codes are not interchangeable.

Procedure type	CPT® code	Description
Procedure examples, not interchangeable	96450	Chemotherapy administration, into central nervous system (CNS) (eg, intrathecal), requiring spinal puncture
	62272	Spinal puncture, therapeutic, for drainage of cerebrospinal fluid (by needle or catheter)
	62328	Spinal puncture, lumbar, diagnostic; with fluoroscopic or CT guidance
	62329	Injection of diagnostic or therapeutic substance into the subarachnoid (intrathecal) space, including needle placement and imaging guidance (fluoroscopy or CT).
	77003	Fluoroscopic guidance and localization of needle or catheter tip for spine or paraspinous diagnostic or therapeutic injection procedures (eg, epidural or subarachnoid)
	76942	Ultrasonic guidance for needle placement (eg, biopsy, aspiration, injection, localization device), imaging supervision and interpretation
	77012	CT guidance for needle placement (eg, biopsy, aspiration, injection, localization device), radiological supervision and interpretation
Anesthesia, only if provided, documented, and separately reportable	00635	Anesthesia for procedures in lumbar region (diagnostic or therapeutic lumbar puncture)
	99100	Add-on anesthesia code for extreme age; cannot be reported alone and must be supported by the primary anesthesia service
Moderate sedation, only if provided, documented, and separately reportable. Moderate sedation code selection depends on patient age, intraservice time, the HCP performing sedation, payer rules, and documentation.	99151	The initial 15 minutes of intraservice time for moderate (conscious) sedation administered by the same HCP performing the procedure for a patient younger than 5 years of age
	99152	The initial 15 minutes of intraservice time for moderate (conscious) sedation administered by the same HCP performing the procedure for a patient 5 years of age or older
	99153	Each additional 15 minutes of intraservice time for moderate sedation; requires a trained observer who assists in monitoring the patient
	99155	The initial 15 minutes of intraservice time for moderate (conscious) sedation administered by an HCP who is not performing the procedure for a patient younger than 5 years of age
	99156	The initial 15 minutes of intraservice time for moderate (conscious) sedation administered by an HCP who is not performing the procedure for a patient 5 years of age or older
	99157	Each additional 15 minutes of moderate (conscious) sedation administered by an HCP who is not performing the procedure
Other services may apply when separately documented	99234	These codes are not routine administration support codes and may apply only when the patient is actually admitted or placed in observation and documentation supports the required level of service.
	99235	These codes are not routine administration support codes and may apply only when the patient is actually admitted or placed in observation and documentation supports the required level of service.
	99236	These codes are not routine administration support codes and may apply only when the patient is actually admitted or placed in observation and documentation supports the required level of service.
Procedure/E/M/ anesthesia modifiers	22	Increased procedural services
	23	Unusual anesthesia
	25	Applies only to a significant, separately identifiable E/M service by the same physician or other qualified healthcare professional on the same day as another procedure or service. It is not a drug-line modifier.
	51	Multiple procedures
	52	Reduced services
	53	Discontinued procedure

CPT=Current Procedural Terminology; CT=computed tomography; E/M=evaluation and management.

Please see additional Important Safety Information on page 12 and the full Prescribing Information, including Boxed WARNING.

Sample Coding for Lembaz™ (adrabetadex) (cont'd)

Code Bundling[8,10-12]*

Certain procedure and service codes may be bundled under payer-specific coding and reimbursement rules. Follow applicable NCCI PTP edits and payer-specific requirements when determining which codes to report.

	CPT® codes	Rules
Codes reportable together	62328 or 62329 + 00635	• Different services furnished by different providers • No modifier needed
	62328 or 62329 + 99100	• 99100 is a qualifying circumstance add-on code for anesthesia when criteria are met • Must accompany an anesthesia code, such as 00635
	62328 or 62329 + 99234/99235/99236	• Only if the E/M service is separately identifiable and medically necessary • Needs modifier -25 when appropriate
Commonly bundled codes that are not separately reportable or reimbursed	62328 or 62329 + 76942	• 62328 and 62329 already include imaging guidance • Separate ultrasound guidance is typically not payable
	62328 or 62329 + 77003	• Fluoroscopic guidance is included in 62328 and 62329
	62328 or 62329 + 77012	• CT guidance is included in 62328
	62328 or 62329 + 62272	• If CSF is withdrawn during the same lumbar puncture used for drug administration, Medicare may view the drainage as integral to the injection procedure • Separate billing is strongest only when documentation supports a distinct therapeutic drainage service
	62328 or 62329 + 96450	• 96450 describes intrathecal chemotherapy administration • Not appropriate to report both for the same injection event
Moderate sedation codes	99153 99157	The moderate sedation codes have parent/add-on relationships with the second code only being required based on time. There are codes that are not reportable alone because they are add-on codes
Codes that may be payable but require a modifier and documentation	99234/99235/99236 with 62328 or 62329	• May be payable only if a significant and separately identifiable E/M service • Must prove medical necessity beyond the procedure itself • Proper modifier used
	62272 with 62328 or 62329	• CSF withdrawal before drug administration is often considered part of the administration process • Documentation must support a distinct therapeutic drainage purpose

For more information about proper coding and bundling, visit CMS.gov.

CSF=cerebrospinal fluid; CT=computed tomography; E/M=evaluation and management; NCCI PTP=National Correct Coding Initiative Procedure-to-Procedure.

*This code bundling list is not all-inclusive; appropriate coding can vary by patient, setting of care, and payer. Correct coding is the responsibility of the provider submitting the claim for the item or service. Please check with the payer to verify codes and special billing requirements.

Sample Coding for Lembaz™ (adrabetadex) (cont'd)

Revenue code examples for outpatient hospital-based facilities[13]*

Revenue codes apply to institutional UB-04/837I billing and should not be used on professional CMS-1500/837P claims. Facilities should validate revenue-code selection through chargemaster, revenue-cycle, payer, MAC, and institutional policy review. Use revenue code examples only as applicable.

Revenue code	Description
0710	Use only as applicable; not a routine drug-administration revenue code. Recovery room services, if separately provided, documented, and billable
0369	Use only as applicable. Operating room services, other
0490	Use only as applicable. Ambulatory surgical care
0370	Use only when anesthesia services are provided, documented, and billable. Anesthesia services, if provided, documented, and billable
0636	Candidate drug/pharmacy revenue code when payer and facility policy support it. Drug/pharmacy line, when payer and facility policy support detailed drug coding
0250	Non-generic drug

ASC=ambulatory surgical center; MAC=Medicare Administrative Contractor.

*Including off-campus clinic, on-campus facility, hospital-based ASC, and other outpatient outlets operated by a hospital.

Sample CMS-1450/UB-04 Claim Form

For outpatient hospital-based facilities*

This form is used by outpatient hospital-based facilities to submit claims to the patient's health plan for facility services, procedures, drugs, and supplies provided. **Codes shown here are an example.** Final billing decisions depend on payer requirements, patient-specific factors, site of care, and current coding guidance.

Field 4

Enter the appropriate type of bill code; for example†

- 013X, Hospital outpatient
- 074X, Clinic OPT
- 083X, Hospital outpatient ASC

†X represents a placeholder for the fourth digit, which indicates the sequence of this bill in particular episode of care (eg, "1" for admit through discharge claim).

Field 42

Enter the appropriate revenue codes and corresponding description of service in field 43; for example:

- **0636**, Drug/pharmacy when facility and payer policy support it
- **0710**, Only for separately billable recovery room
- **0370**, Only for separately billable anesthesia

Field 43

Enter the revenue description. If NDC is required, follow payer/Medicaid/837 format and unit/quantity rules, for example:

- N487230010101ML1 (illustrative only)

Field 44

Enter the applicable HCPCS/CPT codes and modifiers. Drug HCPCS/NOC and procedure CPT lines should be listed separately. **For example:**

- **C9399** or a payer-directed institutional NOC may apply for the drug line
- **62329** Injection of diagnostic or therapeutic substance into the subarachnoid (intrathecal) space, including needle placement and imaging guidance (fluoroscopy or CT)
- **96450** may apply when CNS administration is documented

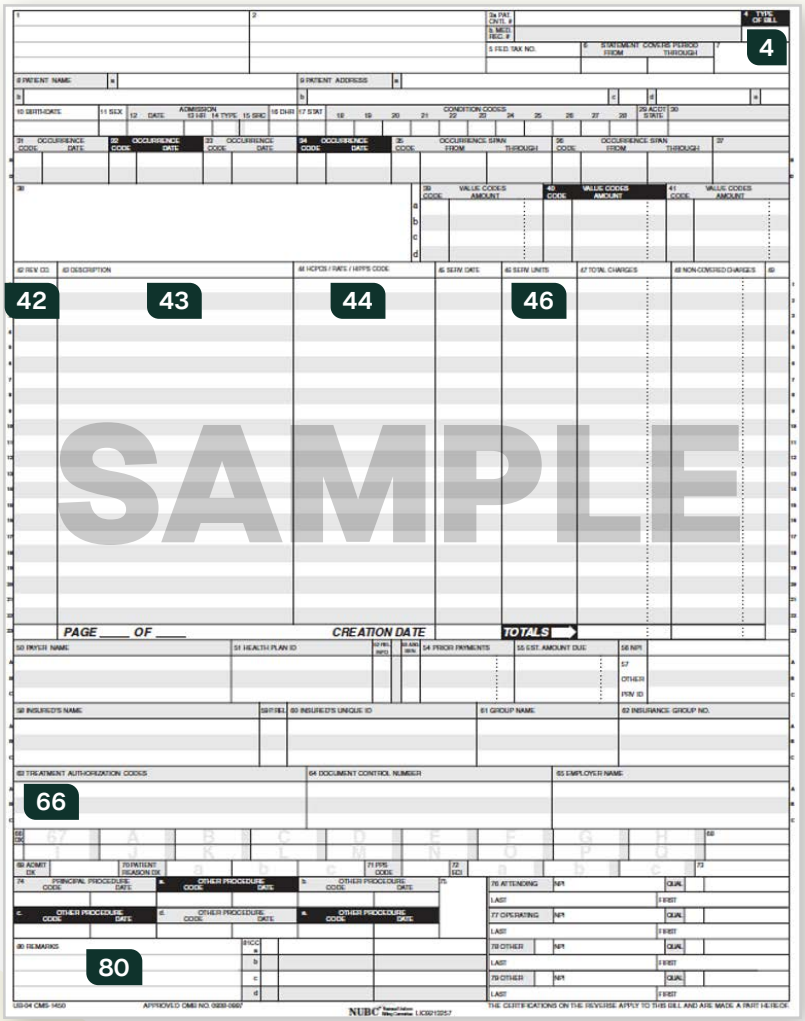
Note: 62270, 62272, and 77003 apply only if separately performed, documented, and reportable. Guidance codes cannot be stacked for the sample needle placement

Modifiers: Use only when payer/site rules support them. For example:

- **C9399TB** or payer-directed NOC with **TB** (illustrative only)

Field 46

Enter the appropriate number of units of service.



Field 66

Enter the appropriate primary ICD-10-CM diagnosis code; for example:

- E75.242, Niemann-Pick disease, type C

Field 80

Enter payer-required drug-identifying information, such as drug name, NDC, dosage, method of administration, when required)

If billing occurs separately, the facility uses CMS-1450 and the HCP uses CMS-1500, resulting in separate claims and reimbursement.

ASC=ambulatory surgical center; CNS=central nervous system; CPT=Current Procedural Terminology; CT=computed tomography; HCPCS=Healthcare Common Procedure Coding System; ICD-10-CM=*International Classification of Diseases, Tenth Revision, Clinical Modification*; NDC=National Drug Code; NOC=Not Otherwise Classified; OPT=outpatient physical therapy.

*Including off-campus clinic, on-campus facility, hospital-based ASC, and other outpatient outlets operated by a hospital.

Please see additional Important Safety Information on page 12 and the full Prescribing Information, including Boxed WARNING.

Sample CMS-1450/UB-04 Claim Form (cont'd)

For inpatient hospital facilities

This form is used by inpatient hospital facilities to submit claims to the patient's health plan for all facility services, including room and board, procedures, medications, and ancillary services provided during a hospital stay. **Codes shown here are an example.** Final billing decisions depend on payer requirements, patient-specific factors, site of care, and current coding guidance.

Field 4

Enter the appropriate type of bill code; for example*

- **011X**, Hospital inpatient
- **014X**, Clinic OPT

*X represents a placeholder for the fourth digit, which indicates the sequence of this bill in particular episode of care (eg, "1" for admit through discharge claim).

Field 42

Enter the appropriate revenue codes and corresponding description of service in field 43; for example:

- **0636**, Drug/pharmacy when facility and payer policy support it
- **0710**, Only for separately billable recovery room
- **0370**, Only for separately billable anesthesia

Field 43

Enter the revenue description. If NDC is required, follow payer/Medicaid/837 format and unit/quantity rules, for example:

- N487230010101ML1 (illustrative only)

Field 44

If detailed institutional line reporting Is required, keep drug HCPCS/NOC and procedure examples on separate lines and validate inpatient facility requirements. **For example:**

- A payer-directed institutional NOC may apply for the drug line, and **96450** may apply only if separately supported by documentation

Modifiers: Use only when payer/site rules support them. For example:

- **C9399TB** or payer-directed NOC with **TB** (illustrative only)

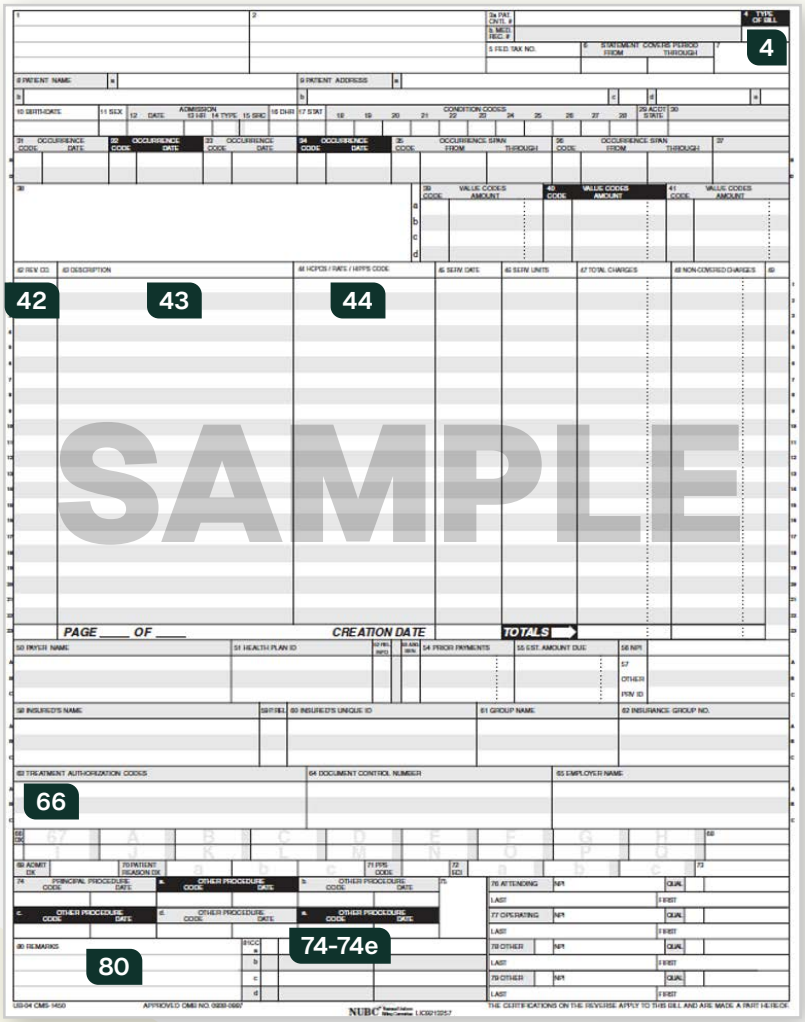
Field 66

Enter the appropriate primary ICD-10-CM diagnosis code; for example:

- E75.242, Niemann-Pick disease, type C

Field 80

Enter payer-required drug-identifying information, such as drug name, NDC, dosage, method of administration, when required



Field 74-74e

Enter the appropriate principal and other ICD-10-PCS procedure codes (along with corresponding dates); for example:

- **009U3ZX**, Drainage of cerebrospinal fluid, spinal canal, percutaneous approach, diagnostic
- **009U3ZZ**, Drainage of cerebrospinal fluid, spinal canal, percutaneous approach

Note: Other ICD-10-PCS procedure codes may apply when supported.

HCPCS=Healthcare Common Procedure Coding System; ICD-10-CM=*International Classification of Diseases, Tenth Revision, Clinical Modification*; ICD-10-PCS=*International Classification of Diseases, Tenth Revision, Procedure Coding System*; NDC=National Drug Code; NOC=Not Otherwise Classified; OPT=outpatient physical therapy.

Please see additional Important Safety Information on page 12 and the full Prescribing Information, including Boxed WARNING.

Sample CMS-1500 Claim Form

For professional services

This form is used to submit claims to the patient's health plan for professional services such as office visits, procedures, and other non-facility medical services provided. **Codes shown here are an example.** Final billing decisions depend on payer requirements, patient-specific factors, site of care, and current coding guidance.

Field 19

Enter additional product information as required by the payer for claims with miscellaneous/unclassified J-code, such as the drug name, 11-digit NDC number, concentration, amount, and route of administration, for example:

- Lembaz 87230-0101-01 carton containing one 900 mg/4.5 mL (200 mg/mL) single-dose vial

Field 21A

Enter the appropriate primary ICD-10-CM diagnosis code; for example:

- E75.242, Niemann-Pick disease, type C

Field 24A

Enter a 6-digit or 8-digit (month-day-year) date for each procedure, service or supply. When NDC is required, report N4 qualifier, 11-digit NDC, and payer-required quantity qualifier in the shaded red box above the date. For example:

- N487230010101ML1 (illustrative only)

Field 24B

Enter the appropriate professional place of service code when applicable; for example:

- **19,** Off-campus outpatient hospital
- **21**, Inpatient hospital
- **22**, On-campus outpatient hospital

Note: These POS examples do not replace institutional UB-04 billing.

Field 24D

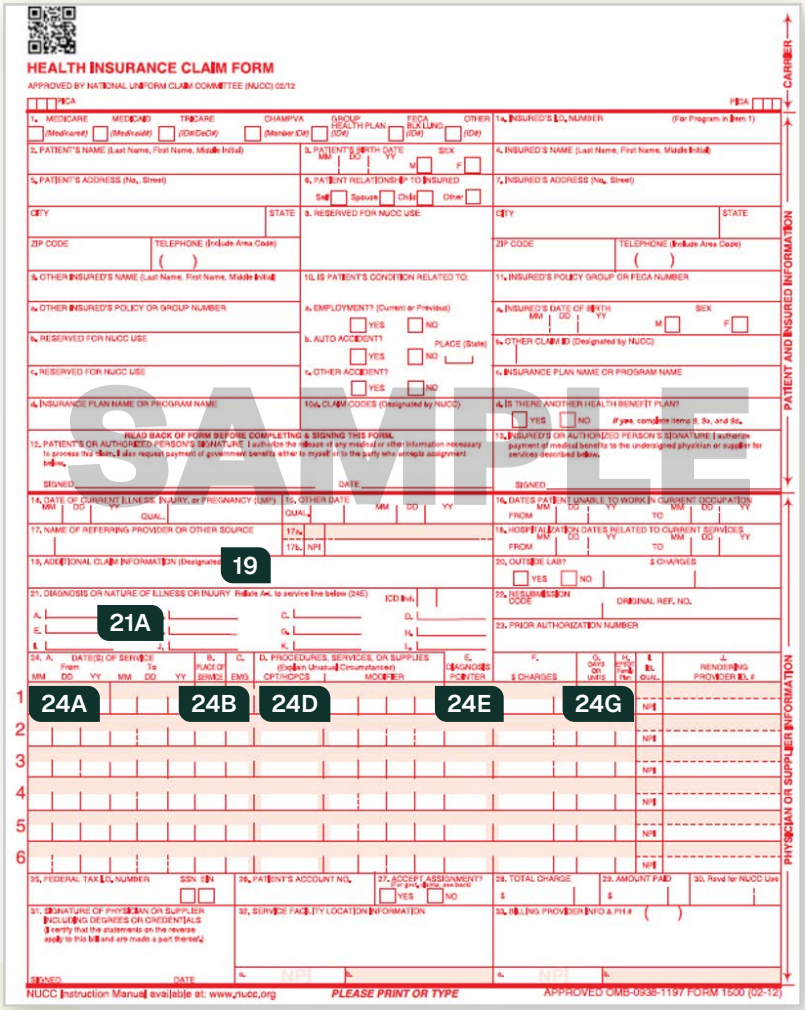
Enter the appropriate CPT/HCPCS codes; for example:

- **96450**, Chemotherapy may apply for CNS administration (eg, intrathecal), requiring and including spinal puncture
- **62329**, Injection of diagnostic or therapeutic substance into the subarachnoid (intrathecal) space, including needle placement and imaging guidance (fluoroscopy or CT)
- **62272**, Spinal puncture, therapeutic, for drainage of cerebrospinal fluid (by needle or catheter)

Note: 62270 and **62272** should only be reported if separately performed, documented, and separately reportable.

Modifiers may apply; for example:

- **JZ** applies to the drug line when required and indicates zero drug wasted/no discarded amount. It should not be administered to any patient



Field 24E

Enter the diagnosis code reference number or letter (as appropriate, per form version) as shown in Field 21A to relate the date of service and the procedures performed to the primary diagnosis.

Field 24G

Enter the appropriate number of units of service.

If billing occurs separately, the HCP uses CMS-1500 and the facility uses CMS-1450, resulting in separate claims and reimbursement.

CNS=central nervous system; CPT®=Current Procedural Terminology; CT=computed tomography; HCPCS=Healthcare Common Procedure Coding System; ICD-10-CM=*International Classification of Diseases, Tenth Revision, Clinical Modification*; NDC=National Drug Code; POS=point of sale.

Please see additional Important Safety Information on page 12 and the full Prescribing Information, including Boxed WARNING.

Sample CMS-1500 Claim Form (cont'd)

For HCP offices

This form is used to submit claims to the patient's health plan for professional services, including office visits, procedures, and other outpatient care provided in the physician office setting. **Codes shown here are an example.** Final billing decisions depend on payer requirements, patient-specific factors, site of care, and current coding guidance.

Field 19

Enter additional product information as required by the payer for claims with miscellaneous/unclassified J-code, such as the drug name, 11-digit NDC number, concentration, amount, and route of administration, for example:

- Lembaz 87230-0101-01 carton containing one 900 mg/4.5 mL (200 mg/mL) single-dose vial

Field 21A

Enter the appropriate primary ICD-10-CM diagnosis code; for example:

- E75.242, Niemann-Pick disease, type C

Field 24A

Enter a 6-digit or 8-digit (month-day-year) date for each procedure, service or supply. When NDC is required, report N4 qualifier, 11-digit NDC, and payer-required quantity qualifier in the shaded red box above the date. For example:

- N487230010101ML1 (illustrative only)

Field 24B

Enter the appropriate place of service code; for example:

- **11,** Office

Field 24D

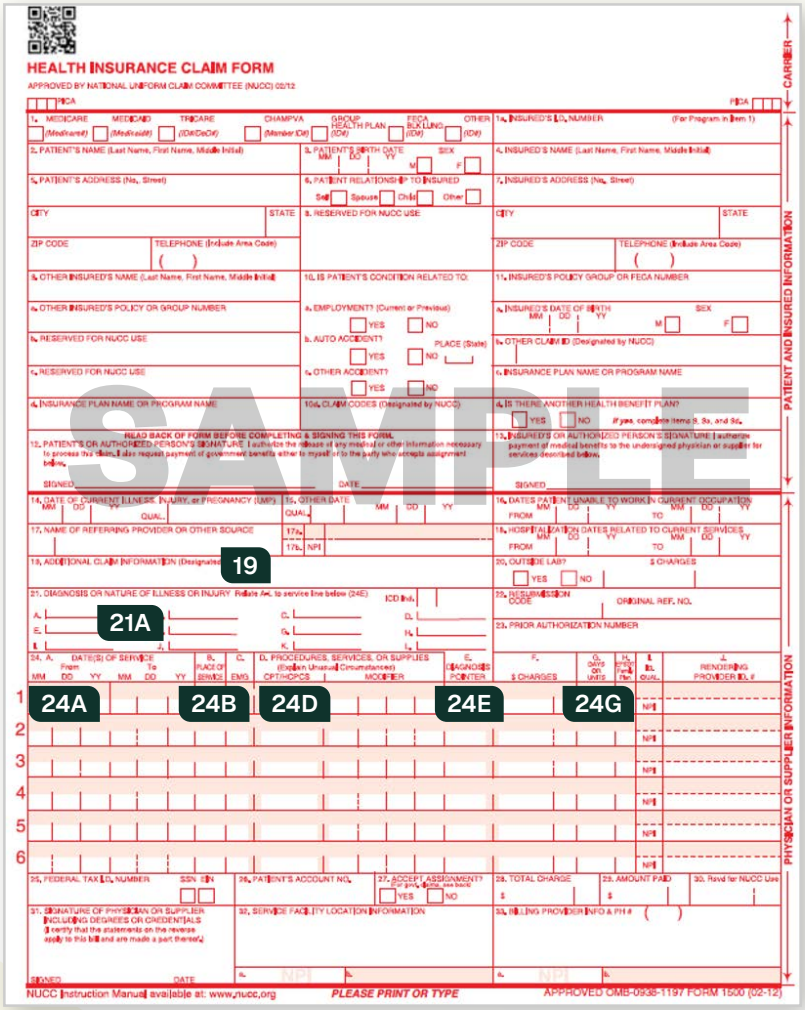
Enter the appropriate CPT/HCPCS codes; for example:

- **96450**, Chemotherapy may apply for CNS administration (eg, intrathecal), requiring and including spinal puncture
- **62270**, Spinal puncture, lumbar, diagnostic
- **62272**, Spinal puncture, therapeutic, for drainage of cerebrospinal fluid (by needle or catheter)

Note: 62270 and **62272** should only be reported if separately performed, documented, and separately reportable.

Modifiers may apply; for example:

- **JZ** applies to the drug line when required and indicates zero drug wasted/no discarded amount. It should not be administered to any patient



Field 24E

Enter the diagnosis code reference number or letter (as appropriate, per form version) as shown in Field 21A to relate the date of service and the procedures performed to the primary diagnosis.

Field 24G

Enter the appropriate number of units of service.

CNS=central nervous system; 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.

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Claim Submission and Tracking Checklists

Completing timely and accurate claims can help facilitate prompt payment. To help proactively prevent denials and underpayment, it is important to review claims before submitting them to a health plan.



Confirm health plan requirements

During the benefits investigation process, confirm that you have identified the following:

- ☑ Coverage and any PA restrictions
- ☑ Billing and coding guidelines
- ☑ Required medical documentation



Check claim for accuracy and completeness

When filling out the claim form, please double check the following:

- ☑ Patient information (eg, patient name, insurer, subscriber name, date of birth, member ID)
- ☑ Provider information (eg, NPI number, name, address, place of service)
- ☑ Coding (eg, ICD-10-CM, CPT®, revenue, and/or HCPCS codes along with appropriate modifiers)
- ☑ Billing units (consistent with the descriptors for the reported CPT® and/or HCPCS codes)
- ☑ Additional information required by the health plan (eg, PA, tax ID, and/or drug NDC number)
- ☑ (If clinical documentation is required) Confirm with the health plan how documentation should be submitted with the initial claim submission



Confirm compliance with claim Submission rules

When submitting the claim, be mindful of the following:

- ☑ Required standards for electronic claims
- ☑ Punctuation and character limit requirements
- ☑ Time frame for submitting claims

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; NPI=National Provider Identifier; PA=prior authorization.

Please see additional Important Safety Information on page 12 and the full Prescribing Information, including Boxed WARNING.

Important Safety Information

WARNING: HEARING IMPAIRMENT INCLUDING PERMANENT HEARING LOSS

Treatment with LEMBAZ has resulted in permanent hearing impairment.

- **Prior to LEMBAZ use, perform a comprehensive audiology evaluation (e.g., pure tone audiometry, speech recognition, tympanometry) to establish baseline hearing levels.**
- **During LEMBAZ treatment, modify LEMBAZ dosage based on comprehensive audiology evaluations as follows:**
  - **To determine the subsequent dosage level, repeat comprehensive audiology evaluation 1 to 2 weeks after the first LEMBAZ dose at each dosage level but before the next dose, and then 1 to 2 weeks after the second LEMBAZ dose at each dosage level but before the next dose.**
  - **When a patient remains at the same LEMBAZ dosage level for 2 consecutive doses or has reached the maximum dosage, perform comprehensive audiology evaluations at one-month intervals until 3 consecutive, stable audiology results are obtained, and then repeat the comprehensive audiology evaluation every 12 months.**

WARNINGS AND PRECAUTIONS

Hearing Impairment Including Permanent Hearing Loss: LEMBAZ-treated patients experienced permanent hearing impairment that ranged from mild to profound hearing loss, including in some patients with normal hearing at baseline. Do not increase the dosage if audiology results show hearing impairment below 4,000 Hz that exceeds the thresholds defined in the full Prescribing Information. In patients with LEMBAZ-associated hearing impairment, data are insufficient to determine whether continuing or lowering the dose affects the risk of further hearing impairment.

Ataxia: LEMBAZ can cause ataxia, including fall, abnormal gait and/or coordination, dizziness, and hypotonia. This may occur shortly after a dose, typically in the first few days. Severe ataxia requiring hospitalization has occurred. Ataxia may be more severe at higher doses. Monitor patients for ataxia severity and consider reducing the dosage to reduce ataxia severity.

Fatigue: LEMBAZ can cause fatigue, including asthenia, fatigue, malaise, and somnolence. This may occur shortly after a dose, typically in the first few days. Severe fatigue requiring hospitalization has occurred. Fatigue may be more severe at higher doses. Monitor patients for fatigue severity and consider reducing the dosage to reduce fatigue severity.

ADVERSE REACTIONS

The most common adverse reactions (reported in ≥25% of patients) were hearing impairment, ataxia, fatigue, vomiting, back pain, headache, pyrexia, diarrhea, nausea, and cough.

To report SUSPECTED ADVERSE REACTIONS, contact Mandos, LLC at 1-844-237-3600 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

Administer LEMBAZ intrathecally by, or under the supervision of, a healthcare provider experienced in performing lumbar punctures.

Please see full Prescribing Information, including Boxed WARNING.

IF YOU HAVE QUESTIONS

844-237-3600

855-595-2575

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Please see additional Important Safety Information on page 12 and the full Prescribing Information, including Boxed WARNING.

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PRO-1247-US-EN-v4, November 2026

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