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

Steps to Help Secure Access to Lembaz™ (adrabetadex)

Use this checklist to help you navigate the steps to secure access to Lembaz.

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.**

STEP 1 | Complete and submit the Patient Enrollment Form (Start Form)

- ☐ Complete the Start Form to begin requesting Lembaz and connect with your Access & Reimbursement Manager (ARM)
- ☐ Confirm the patient's benefits and the health plan's requirements for the prior authorization (PA), treatment location, coding, billing, and patient costs
- ☐ Have your patient provide consent by completing and submitting the Patient Consent & Authorization Form to enroll in Lembaz™ Family Support

Please see additional Important Safety Information on page 4 and the full Prescribing Information, including Boxed WARNING.

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STEP 2 | Complete and submit the PA request

If you need help with the PA, contact your ARM.

- [ ] Fill out the appropriate PA form and include payer-required and supporting documentation, such as:
  - If appropriate, request expedited review by writing on the top of the PA form
  - Patient information
    - Patient name, date of birth, insurance policy number, and diagnosis of infantile-onset Niemann-Pick disease, type C (I-NPC), including the ICD-10-CM code
  - Prescriber information
    - Physician name, NPI
  - Site of care and treatment information
    - Facility name, setting of care, NPI, anticipated date of service, and planned intrathecal administration once every 2 weeks
  - Product and coding information
    - Relevant procedure and HCPCS codes
    - Lembaz injection, 900 mg/4.5 mL (200 mg/mL), single-dose vial; NDC: 87230-101-01[1]
  - Treatment plan
    - Patient-specific dose and titration plan: the starting dose is 100 mg every two weeks for patients younger than 6 months, 200 mg every two weeks for patients aged 6 months to younger than 4 years, and 400 mg every two weeks for patients 4 years or older; the maximum dose is based on weight and tolerability[1]
    - Routine audiologic evaluations and monitoring for fatigue and ataxia during treatment[1]
  - Supporting documentation
    - Letter of medical necessity (please ask your ARM for the template), including the provider ID
    - Relevant clinical documentation, including a complete baseline audiology evaluation before treatment
    - Documentation of prior Expanded Access Program (EAP) participation, if applicable
    - Previous or current treatments and therapies
    - Patient-specific clinical notes documenting I-NPC and the neurological manifestations to be treated
    - Relevant laboratory results
    - Current FDA-approved U.S. Prescribing Information
    - Genetic confirmation
- [ ] Check whether the health plan requires specific assessments or records completed within a set timeframe
- [ ] Confirm that the request lists the planned dose, number of vials or billing units, dosing frequency, NDC, and any procedure or HCPCS codes required by the plan
- [ ] Submit the PA form and supporting documentation using the health plan's preferred method

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STEP 3 | Track the status of the PA request

- [ ] Maintain a log of the PA submissions and dates and methods of correspondence from the payer
- [ ] Confirm that the health plan received the request and ask whether anything else is needed
- [ ] Keep track of names and titles of insurance representatives and reviewers whom you speak with
- [ ] Follow up as needed and provide any requested documentation as soon as possible
- [ ] When approval is received, confirm that the authorized drug, dose, frequency, site of care, and effective dates match the request
- [ ] Record the authorization number, how Lembaz will be obtained, and any renewal requirements

If the PA is approved, go to step 4. If the PA is denied, go to step 5.

STEP 4 | Order Lembaz and prepare for treatment

Lembaz is exclusively available through CuraScript SD by Evernorth

- [ ] Contact CuraScript SD by Evernorth to begin ordering Lembaz


877-900-9223


Wholesalefax@curascript.com


866-628-8942


Curascriptsd.com

STEP 5 | If the PA is denied

- [ ] Appeal the denial
  - Review the denial letter to understand what needs to be addressed in the appeal letter and where and when to submit the appeal
  - Write an appeal letter. If you need help navigating the denial and appeals process, contact the ARM
  - If appropriate, seek the quickest path to the highest level of appeal
- [ ] Follow up with the health plan if you have not received a decision within the timeframe stated in the denial notice or plan materials
  - Confirm that the appeal was received and the status of the appeal
  - If the denial was upheld, you can ask for a one-time exception or consider filing a complaint with the state's insurance commissioner[2]
    - If the denial is upheld, your patient may be eligible for an external review by an independent third party.[2] Requirements and deadlines vary by plan and state, so follow the instructions in the denial notice and the health plan's appeal process

Lembaz™ Family Support and the ARM are here to support you while you navigate coverage for your patients

Call us at 844-237-3600

Visit **LembazRx.com/HCP** for additional resources and tools to support access to Lembaz

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  - **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.

References: 1. Lembaz Prescribing Information. Beren Therapeutics P.B.C. **2.** National Association of Insurance Commissioners. Health insurance claim denied? How to appeal the denial. Published August 17, 2020. Accessed August 18, 2026. https://content.naic.org/article/consumer-insight-health-insurance-claim-denied-how-appeal-denial

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