OPTIMA HEALTH PLAN

MEDICAL PRIOR AUTHORIZATION/STEP-EDIT REQUEST*

Directions: **The prescribing physician must sign and clearly print name (preprinted stamps not valid) on this request**. All other information may be filled in by office staff; **fax to 1-844-668-1550**. No additional phone calls will be necessary if all information (including phone and fax #s) on this form is correct. **If information provided is not complete, correct, or legible, authorization can be delayed**.

For Medicare Members: Medicare Coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: https://www.cms.gov/medicare-coverage-database/overview-and-quick-search.aspx. Additional indications may be covered at the discretion of the health plan.

Drug Requested: Xenpozyme™ (olipudase alfa) **(J0218) (Medical)**

MEMBER & PRESCRIBER INFORMATION: Authorization may be delayed if incomplete.

Member Name: _____

Member Optima #: _____ **Date of Birth:** _____

Prescriber Name: _____

Prescriber Signature: _____ **Date:** _____

Office Contact Name: _____

Phone Number: _____ **Fax Number:** _____

DEA OR NPI #: _____

DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Form/Strength: _____

Dosing Schedule: _____ **Length of Therapy**: _____

Diagnosis: _____ **ICD Code, if applicable:** _____

Weight: _____ **Date:** _____

☐ Standard Review. In checking this box, the timeframe does not jeopardize the life or health of the member or the member's ability to regain maximum function and would not subject the member to severe pain.

Dosing Limits

A. Billable units [NDC Unit]:

- **Xenpozyme 20 mg lyophilized powder for reconstitution in a single-dose vial: 58468-0050-01**
  - **One (1) Xenpozyme 20 mg single-dose vial = 20 billable units**
- **Xenpozyme 4 mg lyophilized powder for reconstitution in a single-dose vial: 58468-0051-01**
  - **One (1) Xenpozyme 4 mg single-dose vial = 4 billable units**

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B. Max Units (per dose and over time) [HCPCS Unit]:

- **Recommended maintenance dose: 3 mg/kg via intravenous infusion every 2 weeks**
- ***Use actual body weight for patients with a BMI ≤ 30. For patients with a BMI > 30, calculate adjusted body weight (kg) = (actual height in m)^2× 30**

Xenpozyme (olipudase alfa), a hydrolytic lysosomal sphingomyelin-specific enzyme approved by the Food and Drug Administration (FDA) for treatment of non–central nervous system manifestations of acid sphingomyelinase deficiency (ASMD) in adult and pediatric individuals.

Xenpozyme is an intravenous enzyme replacement therapy and the first FDA-approved treatment for ASMD. Xenpozyme is not expected to cross the blood-brain barrier and is intended to treat the non-central nervous system disease manifestations. Xenpozyme has only been studied in individuals with the type B and A/B phenotypes.

Xenpozyme has a black box warning for severe hypersensitivity reactions including anaphylaxis. Appropriate medical support measures, including cardiopulmonary resuscitation equipment, should be readily available during Xenpozyme administration. If a severe hypersensitivity reaction occurs, Xenpozyme should be discontinued immediately, and appropriate medical treatment should be initiated.

CLINICAL CRITERIA: Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

Initial Authorization: 12 months

- ❑ Member has a diagnosis of acid sphingomyelinase deficiency (ASMD) confirmed by **ONE** of the following:
  - ❑ Pathogenic sphingomyelin phosphodiesterase-1 (SMPD1) gene mutation
  - ❑ Deficiency in acid sphingomyelinase (ASM) enzyme activity <10% of controls as measured in fibroblasts, leukocytes or dried blood spot **[Note: NPD-A (infantile neurovisceral ASMD) has not been studied. Genotype-phenotype correlations as well as signs/symptoms may not be conclusive in infants therefore requests will be evaluated on a case-by-case basis]**
- ❑ Member has a clinical presentation consistent with **ONE** of the following ASMD types:
  - ❑ ICD Code: E75.241 Niemann-Pick type B
  - ❑ ICD Code: E75.244 Niemann-Pick type A/B
- ❑ Requested medication will be used for the treatment of non-central nervous system disease manifestations
- ❑ Member should **NOT** require invasive ventilatory support **OR** non-invasive ventilatory support while awake and for >12 hours a day **(Note: Members requiring ventilatory support will be reviewed on a case-by-case basis)**

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- ❑ Provider must submit documentation for at least **ONE** of the following:
  - ❑ Baseline: percent predicted diffusion capacity of the lungs for carbon monoxide (DLco) or age- appropriate pulmonary function testing
  - ❑ Baseline: spleen and/or liver volumes
  - ❑ Baseline: plasma lyso-sphingomyelin
  - ❑ Baseline: Platelet count
  - ❑ Baseline: height Z-score and skeletal maturation (relevant for pediatric patients)
- ❑ Provider has submitted baseline transaminase (alanine aminotransferase [ALT] and aspartate aminotransferase [AST]) levels obtained within 1 month prior to treatment initiation
- ❑ Females of reproductive potential will have pregnancy status verified prior to start of therapy and will use effective contraception during treatment and for 14 days after the last dose if therapy is discontinued
- ❑ Requested dose has been provided: _____

Reauthorization: 12 months. Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

- ❑ Member has experienced an absence of unacceptable toxicity from the drug (e.g., anaphylaxis and severe hypersensitivity reactions, severe infusion-associated reactions, severely elevated liver transaminases)
- ❑ Member has **NOT** experienced progressive/irreversible severe cognitive impairment
- ❑ Member has shown disease improvement or stability from pre-treatment baseline as demonstrated by at least **ONE** of the following:
  - ❑ Improvement in or stability in the percent predicted diffusion capacity of the lungs for carbon monoxide (DLco) or other age-appropriate pulmonary function testing
  - ❑ Improvement in or stability of spleen and/or liver volumes
  - ❑ Reduction in plasma lyso-sphingomyelin
  - ❑ Improvement in or stability of platelet count
  - ❑ Improvement in linear growth progression as measured by mean height Z-scores (pediatric patients only)
- ❑ Provider has submitted transaminase (alanine aminotransferase [ALT] and aspartate aminotransferase [AST]) levels obtained within the last 30 days
- ❑ Requested dose has been provided: _____

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Medication being provided by: Please check applicable box below.

- ❑ **Location/site of drug administration:** _____
  NPI or DEA # of administering location: _____

OR

- ❑ **Specialty Pharmacy – Proprium Rx**

For urgent reviews: Practitioner should call Optima Pre-Authorization Department if they believe a standard review would subject the member to adverse health consequences. Optima's definition of urgent is a lack of treatment that could seriously jeopardize the life or health of the member or the member's ability to regain maximum function.

Use of samples to initiate therapy does not meet step edit/ preauthorization criteria.

Previous therapies will be verified through pharmacy paid claims or submitted chart notes.