Division: Pharmacy Policy	Subject: Prior Authorization Criteria
Original Development Date: Original Effective Date: Revision Date:	November 15, 2022

Xenpozyme™ (olipudase alfa-rpcp)

LENGTH OF AUTHORIZATION: Initial: 6 months
Continuation: 1 Year

REVIEW CRITERIA:

- Patient must have a diagnosis of acid sphingomyelinase deficiency (ASMD) with non-central nervous system manifestations (e.g., Hepatosplenomegaly, interstitial lung disease, etc.).
- Documentation of one of the following must be provided confirming the diagnosis:
  - Genetic test results confirming the presence of pathogenic variants in the sphingomyelin phosphodiesterase-1 (SMPD1) gene.
  - Peripheral blood leukocytes or cultured skin fibroblasts test results confirming a decrease in residual acid sphingomyelinase (ASM) activity.
- Baseline transaminase levels (alanine aminotransferase [ALT] and aspartate aminotransferase [AST]) drawn within 1 month prior to the initiation of therapy. *(Official labs must be provided)*
- Females of reproductive potential should have a negative pregnancy test prior to the initiation of therapy.
- Must be prescribed by, or in consultation with a hepatologist, pulmonologist, or related specialist with expertise in the diagnosis and management of ASMD.

CONTINUATION OF THERAPY

- Patient met initial review criteria.
- Documentation of improved clinical response (e.g., Increased predicted diffusing capacity for carbon monoxide (DLco), increased platelet count, reduction in liver volume, reduction in spleen volume).
- Patient has NOT experienced serious treatment-related adverse events.
- Dosing is appropriate as per labeling or is supported by compendia.

DOSING AND ADMINISTRATION:

- Refer to product labeling at https://www.accessdata.fda.gov/scripts/cder/daf/
- Available as 20mg lyophilized powder in a single-dose vial.