Division: Pharmacy Policy	Subject: Prior Authorization Criteria
Original Development Date: Original Effective Date: Revision Date:	December 8, 2021 May 8, 2024

Pompe Disease Agents

Preferred agents: N/A

Non-preferred agents: Lumizyme® (alglucosidase alfa), Nexviazyme™ (avalglucosidase alfa-ngpt), Opfolda™ (miglustat) and Pombiliti™ (cipaglucosidase alfa-atga)

LENGTH OF AUTHORIZATION: Up to 1 year

INITIAL REVIEW CRITERIA:

- Patient must have a diagnosis of Pompe Disease (lysosomal acid alpha-glucosidase [GAA] deficiency).
- Patient must be ≥ 1 year of age for Nexviazyme.
- Patients of all ages can be prescribed Lumizyme.
- For Pombiliti™ and Opfolda™, all the following must be met:
  - Patient must be ≥ 18 years of age and weigh ≥ 40 kg.
  - Patient had an inadequate response or intolerance to current enzyme replacement therapy (ERT) (e.g., Lumizyme® /alglucosidase alfa, Nexviazyme™ /avalglucosidase alfa-ngpt). (*Clinical documentation demonstrating response to previous therapy must be submitted).*
  - Pombiliti™ and Opfolda™ will be administered as combination therapy.

CONTINUATION OF THERAPY:

- Patient met initial review criteria.
- Documentation of improved clinical response.
- 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/