HIGHMARK WHOLECARE

Updated: 09/2022
PARP Approved: 10/2022

Prior Authorization Criteria

Enzyme Replacement Therapy, Pompe Disease

All requests for Enzyme Replacement Therapy for Pompe Disease require a prior authorization and will be screened for medical necessity and appropriateness using the criteria listed below.

Enzyme Replacement Therapy for Pompe Disease medications include Lumizyme (alglucosidase alfa) and Nexviazyme (avalglucosidase alfa-ngpt). New products with this classification will require the same documentation.

Enzyme Replacement Therapy for Pompe Disease Prior Authorization Criteria:

Coverage may be provided with a diagnosis of **Pompe Disease** and the following criteria is met:

- Must be age-appropriate according to FDA-approved labeling, nationally recognized compendia, or evidence-based practice guidelines
    - For Nexviazyme (avalglucosidase alfa-ngpt), must be at least 1 year of age.
- Medication must be prescribed by or in consultation with a metabolic specialist and/or biochemical geneticist.
- Must have GAA assay performed on dried blood spots, skin fibroblasts or muscle biopsy.
- For members 12 years and older, must have pulmonary function testing (PFT) and muscle strength evaluation with documentation of baseline percent predicted forced vital capacity (FVC) and baseline 6-minute walk test.
- For late-onset Pompe disease only, must have completed genetic testing to identify the specific mutation to confirm the diagnosis of late-onset Pompe disease.
- **Initial Duration of Approval:** 12 months
- **Reauthorization criteria**
    - For members 12 years of age or older, clinical documentation of improvement defined by improvement in percent predicted FVC and/or 6-minute walk test compared to baseline.
    - For members under the age of 12, reauthorization benefit will be approved if there is documented, significant improvement with prior courses of treatment.
- **Reauthorization Duration of approval:** 12 months

Coverage may be provided for any non-FDA labeled indication if it is determined that the use is a medically accepted indication supported by nationally recognized pharmacy compendia or peer-reviewed medical literature for treatment of the diagnosis(es) for which it is prescribed. These requests will be reviewed on a case by case basis to determine medical necessity.

When criteria are not met, the request will be forwarded to a Medical Director for review. The physician reviewer must override criteria when, in their professional judgment, the requested medication is medically necessary.

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ENZYME REPLACEMENT THERAPY, POMPE DISEASE
PRIOR AUTHORIZATION FORM

Please complete and fax all requested information below including any progress notes, laboratory test results, or chart documentation as applicable to Highmark Wholecare Pharmacy Services. **FAX:** (888) 245-2049
If needed, you may call to speak to a Pharmacy Services Representative. **PHONE**: (800) 392-1147 Mon – Fri 8:30am to 5:00pm

PROVIDER INFORMATION

Requesting Provider:	Provider NPI:
Provider Specialty:	Office Contact:
State license #:	Office NPI:
Office Address:	Office Phone:
	Office Fax:

MEMBER INFORMATION

Member Name:	DOB:	
Member ID:	Member weight:	Height:

REQUESTED DRUG INFORMATION

Medication:	Strength:	
Directions:	Quantity:	Refills:
Is the member currently receiving requested medication? ☐ Yes ☐ No	Date Medication Initiated:	

Billing Information

This medication will be billed: ☐ at a pharmacy **OR** ☐ medically, JCODE:_____
Place of Service: ☐ Hospital ☐ Provider's office ☐ Member's home ☐ Other

Place of Service Information

Name:	NPI:
Address:	Phone:

MEDICAL HISTORY (Complete for ALL requests)

Diagnosis: ☐ Infantile-onset Pompe disease ☐ Late-onset (non-infantile) Pompe disease ICD-10: _____
☐ Other: _____ ICD-10: _____
Was diagnosis confirmed by GAA assay performed on dried blood spots, skin fibroblasts or muscle biopsy? ☐ Yes ☐ No
For late-onset Pompe disease, was genetic testing done to confirm the diagnosis? ☐ Yes ☐ No
Has baseline pulmonary function testing (PFT) and muscle strength evaluation been completed? ☐Yes ☐ No

CURRENT or PREVIOUS THERAPY

Medication Name	Strength/ Frequency	Dates of Therapy	Status (Discontinued & Why/Current)

REAUTHORIZATION

Please indicate which of the following apply as a result of treatment:

☐ Improvement in FVC:
Baseline: _____ Date:_____
Recent FVC: _____ Date: _____

☐ Improvement in 6-min walk or other muscle strength evaluation:
Baseline: _____ Date: _____
Recent 6-min walk: _____ Date: _____

☐ Other improvements (please describe or attach chart documentation): _____

SUPPORTING INFORMATION or CLINICAL RATIONALE

Prescribing Provider Signature	Date