SENTARA COMMUNITY PLAN
(MEDICAID)

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 1-844-305-2331**. 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**.

Drug Requested: (select one drug below)

❑ **Nexviazyme®** (avalglucosidase alfa-ngpt) **(J0219)**	❑ **Pombiliti™** (cipaglucosidase alfa-atga) + **Opfolda™** (miglustat) **(J1203, J1202)**

Provider please select requested access for Opfolda™(miglustat):

- ❑ Pharmacy Benefit Requests can also be submitted using the following form: **Opfolda**
- ❑ Medical Buy & Bill

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

Member Name: _____

Member Sentara #: _____ **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 Name/Form/Strength: _____

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

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

Weight: _____ **Date:** _____

PROVIDER PLEASE NOTE: If member meets all clinical requirements for Pombiliti™ and is granted an approval, a subsequent authorization will be entered to allow the member to receive Opfolda™ at the maximum recommended quantity of 8 capsules per 28 days via the pathway selected above.

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Recommended Dosage:

Drug	Dose

Nexviazyme 100 mg vial	• Nexviazyme is administered as intravenous infusion. For patients weighing:
	  ○ ≥30 kg, the recommended dosage is 20 mg/kg (actual body weight) every two weeks
	  ○ <30 kg, the recommended dosage is 40 mg/kg (actual body weight) every two weeks
	• Maximum units: 2300 mg every 14 days
	• Prior to administration, consider pretreating with antihistamines, antipyretics, and/or corticosteroids.

Pombiliti 105 mg vial	• Pombiliti is administered as an intravenous infusion. • Patients must weigh ≥ 40 kg. • Administer 20 mg/kg (of actual body weight) every two weeks as an intravenous infusion over approximately 4 hours. • Maximum units: 2300 mg every 14 days • Pombiliti must be administered in combination with Opfolda. If the Opfolda dose is missed, Pombiliti should not be administered. • Prior to Pombiliti administration, consider pretreating with antihistamines, antipyretics, and/or corticosteroids. If premedication was used with previous enzyme replacement therapy (ERT), prior to Pombiliti administration, pretreat with antihistamines, antipyretics, and/or corticosteroids. • Start Pombiliti in combination with Opfolda two weeks after the last ERT dose. • Initiate Pombiliti ~1 hour after oral administration of Opfolda. If the Pombiliti infusion cannot be started within 3 hours of oral administration of Opfolda, reschedule Pombiliti in combination with Opfolda at least 24 hours after Opfolda was last taken.

Opfolda 65 mg capsules	• Opfolda is administered orally every other week. The recommended dosage is based on actual body weight. For patients weighing:
	  ○ ≥50 kg, the recommended dose is 260 mg (4 capsules)
	  ○ ≥40 kg to <50 kg, the recommended dose is 195 mg (3 capsules)
	• In patients with moderate or severe renal impairment, the recommended dosage is based on actual body weight. For patients weighing:
	  ○ ≥50 kg, the recommended dose is 195 mg (3 capsules)
	  ○ ≥40 kg to <50 kg, the recommended dose is 130 mg (2 capsules)

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 must meet **ONE** of the following age requirements:
  - ❑ For Nexviazyme requests: Member is 1 year of age or older
  - ❑ For Pombiliti requests: Member is 18 years of age or older and weighs at least 40 kg
- ❑ Member has a diagnosis of late-onset (non-infantile) disease

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- ❑ Member has a diagnosis of Pompe disease (acid alpha-glucosidase (GAA) deficiency) confirmed by **ONE** of the following:
  - ❑ Deficiency of acid alpha-glucosidase (GAA) enzyme activity which shows reduced enzyme activity less than 40% of the lab specific normal mean value
  - ❑ Detection of biallelic pathogenic variants in the GAA gene by molecular genetic testing
- ❑ Member has documented baseline values for percent predicted forced vital capacity (FVC) and/or 6 minute walk test (6MWT) **(please submit labs)**
- ❑ Member has **NOT** demonstrated an improvement in objective measures (i.e., FVC and 6MWT) after receiving **ONE** of the following for at least one year **(please submit chart notes)**
  - ❑ Lumizyme® (alglucosidase alfa) intravenous infusion
  - ❑ Nexviazyme® (avalglucosidase alfa-ngpt) intravenous infusion
- ❑ Requested medication will **NOT** be used in combination with other enzyme replacement therapies e.g., Lumizyme® (alglucosidase alfa)
- ❑ Members susceptible to fluid volume overload or those with an acute underlying respiratory illness or compromised cardiac or respiratory function, will be closely monitored for exacerbation of their cardiac or respiratory status during infusion
- ❑ For Pombiliti™ requests: Medication will be used in combination with Opfolda™ (miglustat)
  - ❑ Pharmacy Benefit Requests can also be submitted using the following form: **Opfolda**
  - ❑ Medical Buy & Bill

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 continues all initial authorization criteria
- ❑ Member has experienced an absence of unacceptable toxicity from the drug (e.g., anaphylaxis, severe hypersensitivity reactions, severe infusion-associated reactions, acute cardiorespiratory failure)
- ❑ Member has demonstrated a beneficial response to therapy compared to pretreatment baseline in one or more of the following **(please submit labs)**:
  - ❑ Disease stabilization
  - ❑ Improvement in FVC
  - ❑ Improvement in 6MWT
- ❑ Member is being monitored for antibody formation (including neutralizing antibodies)

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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 Sentara Health Plans Pre-Authorization Department if they believe a standard review would subject the member to adverse health consequences. Sentara Health Plan'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.****

***Approved by Pharmacy and Therapeutics Committee**: 11/12/2021
REVISED/UPDATED/REFORMATTED: ~~12/13/2021;~~ ~~12/23/2021;~~ **2/28/2024**