SENTARA HEALTH PLANS

PHARMACY 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 1-800-750-9692**. No additional phone calls will be necessary if all information (including phone and fax #s) on this form is correct. **If the information provided is not complete, correct, or legible, the authorization process may be delayed.**

Gaucher Disease Drugs (Substrate Reduction Therapy)

Drug Requested: (select drug below that applies)

❑ **Cerdelga®** (eliglustat)	❑ **Miglustat** (generic Zavesca®)

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 Form/Strength: _____

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

Diagnosis: _____ **ICD Code:** _____

Weight: _____ **Date:** _____

Note: There is currently insufficient clinical evidence that supports the combination use of substrate reduction therapy with enzyme replacement therapy (e.g., Cerezyme®, Eleyso®, Vpriv®)

Medication	Recommended Dosage
Cerdelga® (eliglustat) • **Note: Dosage is based on patient CYP2D6 metabolizer status (extensive metabolizers [EMs], intermediate metabolizers [IMs], or poor metabolizers [PMs]) determined by an FDA-cleared test.**	EMs and IMs: 84 mg twice daily
	PMs: 84 mg once daily
Miglustat (generic Zavesca®)	100 mg 3 times daily; dose may be reduced to 100 mg 1 to 2 times daily in patients with adverse effects (e.g., tremor, diarrhea)

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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 is 18 years of age or older
- ❑ Prescribed by or in consultation with a metabolic geneticist or physician knowledgeable in the management of Gaucher disease
- ❑ Medication will **NOT** be used in combination with Cerezyme®, Vpriv®, Elelyso®, or other enzyme replacement or substrate-reducing therapy for treatment of Gaucher disease
- ❑ Member has a documented diagnosis of Type I Gaucher Disease as confirmed by **ONE** of the following **(submit documentation)**:
  - ❑ Beta-glucocerebrosidase activity (in leukocytes or skin fibroblasts) of less than 30% of normal values
  - ❑ deoxyribonucleic acid (DNA) testing (mutations in the glucocerebrosidase gene)
- ❑ Member's disease has resulted in at least **ONE** of the following **(Check all that apply; submit labs for baseline criteria)**:
  - ❑ Anemia [i.e., hemoglobin ≤ 11 g/dL (women) or 12 g/dL (men)] not attributed to iron, folic acid, or vitamin B12 deficiency
  - ❑ Moderate to severe hepatomegaly (liver size 1.25 or more times normal volume) or splenomegaly (spleen size 5 or more times normal volume)
  - ❑ Skeletal disease (e.g., lesions, remodeling defects and/or deformity of long bones, osteopenia/osteoporosis)
  - ❑ Symptomatic disease (e.g., bone pain, fatigue, dyspnea, angina, abdominal distension, diminished quality of life)
  - ❑ Thrombocytopenia (platelet count ≤ 120,000/mm^3)
- ❑ Member has tried and failed enzyme replacement therapy or is **NOT** a candidate (e.g., due to allergy, hypersensitivity, or poor venous access) for enzyme replacement therapy (e.g., Cerezyme®, Eleyso®, Vpriv®)
- ❑ For Cerdelga (eligustat) requests only:
  - ❑ CYP2D6 phenotype has been determined by an FDA-cleared test to be **ONE** of the following **(submit labs)**:
    - ❑ Extensive Metabolizer (EM)
    - ❑ Intermediate Metabolizer (IM)
    - ❑ Poor Metabolizer (PM)
  - ❑ Medication may **NOT** be approved for members with any of the following:
    - Pre-existing cardiac conditions (e.g., congestive heart failure, recent acute myocardial infarction, bradycardia, heart block, ventricular arrhythmia, or long QT syndrome)

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- Currently taking class 1A antiarrhythmic medications (e.g., quinidine, procainamide) or Class III antiarrhythmic medications (e.g., amiodarone, sotalol)
- Moderate renal impairment, severe renal impairment, or end-stage renal disease (ESRD)
- Mild, moderate, or severe hepatic impairment or cirrhosis
- Partial or total splenectomy within the last 3 years
- Ultra-rapid or indeterminate CYP2D6 metabolizers
- Type 2 or 3 Gaucher Disease

Reauthorization Approval: 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 is **NOT** on concomitant enzyme replacement therapy
- ❑ Member has experienced disease response with treatment as defined by at least **ONE** of the following compared to pre-treatment baseline **(Check all that apply; submit labs/progress notes)**:
  - ❑ Improvement in symptoms (e.g., bone pain, fatigue, dyspnea, angina, abdominal distension, diminished quality of life)
  - ❑ Reduction in size of liver or spleen
  - ❑ Improvement in hemoglobin/anemia
  - ❑ Improvement in skeletal disease (e.g., increase in lumbar spine and/or femoral neck BMD, no bone crises or bone fractures)
  - ❑ Improvement in platelet counts
- ❑ Member has **NOT** experienced unacceptable toxicity from the drug (e.g., severe diarrhea and weight loss, severe tremors, peripheral neuropathies, thrombocytopenia, ECG changes or cardiac arrhythmias)

Medication being provided by Specialty Pharmacy - PropriumRx

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.