SENTARA HEALTH PLANS

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 1-844-668-1550**. 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**.

For Medicare Members: Medicare Coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: https://www.cms.gov/medicare-coverage-database/overview-and-quick-search.aspx. Additional indications may be covered at the discretion of the health plan.

Drug Requested: Kanuma® (sebelipase alfa) solution for IV Infusion (J2840) (Medical)

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, if applicable:** _____

Weight: _____ **Date:** _____

❑ Standard Review. In checking this box, the timeframe does not jeopardize the life or health of the member or the member's ability to regain maximum function and would not subject the member to severe pain.

Recommended Dosing:

- **Infants, children, adolescents, adult dosing:** 1 mg/kg every other week; may increase to maximum of 3 mg/kg every other week in members with suboptimal response
- **Infant with rapidly progressive disease (within the first 6 months of life):** 1 mg/kg/dose once weekly; may increase to 3 mg/kg/dose once weekly if response not optimal; if response remains not optimal may further increase to maximum of 5 mg/kg/dose once weekly

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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. **(Trials will be verified using pharmacy claims and/or submitted chart notes.)**

Initial Authorization: 6 months.

- ❑ Member is 1 month of age or older and has a diagnosis of Lysosomal Acid Lipase (LAL) deficiency
- ❑ LAL diagnosis has been confirmed by **ONE** of the following:
  - ❑ Biallelic pathogenic variants in LIPA
  - ❑ Deficient LAL enzyme activity in peripheral blood leukocytes, fibroblasts, or dried blood spots
- ❑ Member has at least **ONE** of the following clinical manifestations of LAL deficiency **(check all that apply)**:
  - ❑ Hyperlipidemia despite use of appropriate lipid-lowering therapy
  - ❑ Hepatomegaly
  - ❑ Splenomegaly
  - ❑ Steatosis
  - ❑ Elevated serum transaminases
- ❑ Dose does not exceed 1 mg/kg every other week (1 mg/kg per week for members with rapidly progressive disease presenting within first 6 months of life; may be increased to 3 mg/kg per week upon documentation of suboptimal clinical response to 1 mg/kg per week)
- ❑ Member's current weight must be noted: _____

Reauthorization: 6 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 must continue to meet all initial authorization criteria
- ❑ Member must meet **ONE** of the following:
  - ❑ Member has experienced a positive response to therapy as demonstrated by at least **ONE** of the following **(check all that apply)**:
    - ❑ Improvement in weight-for-age z-scores for members exhibiting growth failure
    - ❑ Improvement in LDL
    - ❑ Improvement in HDL
    - ❑ Improvement in triglycerides
    - ❑ Improvement of AST or ALT

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- ❑ Dose escalation in pediatric and adult members with a suboptimal clinical response to the 1 mg/kg dose defined by at least **ONE** of the following:
  - ❑ Poor growth
  - ❑ Deteriorating biochemical markers [e.g., alanine aminotransferase (ALT), aspartate aminotransferase (AST)], and/or parameters of lipid metabolism [e.g., low-density lipoprotein cholesterol (LDL-c), triglycerides (TG)]
- ❑ Dose escalation for members with rapidly progressive disease presenting within the first 6 months of life who have a suboptimal clinical response to the 1 mg/kg dose or 3 mg/kg dose defined by at least **ONE** of the following:
  - ❑ Poor growth
  - ❑ Deteriorating biochemical markers [e.g., alanine aminotransferase (ALT), aspartate aminotransferase (AST)]
  - ❑ Persistent or worsening organomegaly
- ❑ Member's current weight must be noted: _____

Medication being provided by (check box below that applies):

- ❑ **Location/site of drug administration:** _____

  NPI or DEA # of administering location: _____

OR

- ❑ **Specialty Pharmacy - PropriumRx**

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**: 5/19/2022
REVISED/UPDATED: ~~5/13/2022; 6/3/2022;~~ **6/17/2022**