RX.PA.019.MPC Kanuma® (Sebelipase Alfa)

The purpose of this policy is to define the prior authorization process for Kanuma (sebelipase alfa).

Kanuma (Sebelipase alfa) is indicated for Lysosomal Acid Lypase (LAL) deficiency (Wolman disease) and Cholesteryl Ester Storage Disease (CESD).

The drug, Kanuma (Sebelipase alfa), is subject to the prior authorization process.

PROCEDURE

Initial Authorization Criteria:

Must meet all of the criteria listed under the respective diagnosis:

1. **For all diagnoses:**
   - Must be prescribed by or in consultation with a physician that specializes in the treatment of inherited metabolic disorders
   - Must have confirmation of a genetic defect in the LIPA gene via genetic testing
   - Must not have an allergy or sensitivity to eggs or egg products
   - Must be prescribed at a dose within the manufacturer's dosing guidelines (based on diagnosis, weight, etc) listed in the FDA approved labeling

2. **Rapidly progressive Lysosomal Acid Lypase (LAL) deficiency (Wolman disease) defined as:**
   - Patients within the first 6 months of life
   - Documentation the following:
     - Growth failure with other causes ruled out
     - Other evidence of rapidly progressive disease prior to 6 months of age confirmed via CT scan, MRI, or biopsy
       - Hepatosplenomegaly
       - Ascites
       - Calcification of adrenal gland tissue
       - Liver fibrosis confirmed through biopsy

3. **Cholesteryl Ester Storage Disease (CESD) defined as:**
   - Documentation of the following:
     - LDL-c of ≥130 mg/dL in pediatric patients and ≥160 mg/dL in adult patients
     - Malabsorption with other causes ruled out

    - Growth failure with other causes ruled out
    - Calcification of adrenal gland tissue
        - Confirmed via CT scan or MRI
- Anemia, defined as
    - For members > 12 years of age
        - Hemoglobin < 12 g/dL in males
        - Hemoglobin < 11 g/dL in females
    - For children between 2 and 12 years of age
        - Hemoglobin < 10.5 g/dL
    - For children between 6 months and 2 years of age
        - Hemoglobin < 9.5 g/dL
    - Hepatomegaly
        - Defined as liver size 1.25 or more times normal (Normal liver size is 2.5% of total body weight)
        - Confirmed via CT scan or MRI
    - Splenomegaly
        - Defined as splenic mass greater than normal (Normal spleen size is 0.2% of total body weight)

Reauthorization Criteria:

All prior authorization renewals are reviewed on an annual basis to determine the Medical Necessity for continuation of therapy. Authorization may be extended at 1-year intervals based upon chart documentation from the prescriber that the member's condition has improved based upon the prescriber's assessment while on therapy.

Limitations:

Length of Authorization (if above criteria met)	
Initial Authorization	Up to 1 year
Reauthorization	Same as initial

If the established criteria are not met, the request is referred to a Medical Director for review, if required for the plan and level of request.

HCPCS Code(s):

Code	Description
J2840	Injection, sebelipase alfa, 1 mg

REFERENCES

1. Kanuma [prescribing information]. Cheshire, CT: Alexion Pharmaceuticals; December 2015.
2. Wolman Disease. *National Organization for Rare Disorders (NORD)*.
3. Kruer MC. Lysosomal Storage Disease.Medscape. Updated: December 9, 2015. Available at: http://www.emedicine.com/neuro/topic668.htm Accessed December 15, 2015.
4. Genetics Home Reference. Wolman Disease. October 2007. Available at: http://ghr.nlm.nih.gov/condition=wolmandisease Accessed December 15, 2015.