Policy Title:	Strensiq (asfotase alfa) subcutaneous		
		Department:	PHA
Effective Date:	09/06/2019		
Review Date:	5/7/19, 9/6/2019, 1/29/20, 7/27/2020		
Revision Date:	5/7/19, 9/6/2019, 1/29/20,7/27/2020		

Purpose: To support safe, effective and appropriate use of Strensiq (asfotase alfa).

Scope: Medicaid, Exchange, Medicare-Medicaid Plan (MMP)

Policy Statement:

Strensiq (asfotase alfa) is covered under the Medical Benefit when used within the following guidelines. Use outside of these guidelines may result in non-payment unless approved under an exception process.

Procedure:

Coverage of Strensiq will be reviewed prospectively via the prior authorization process based on criteria below.

Initial Criteria Coverage

- Prescriber is an endocrinologist or specialist in the treatment of perinatal/infantile or juvenile hypophosphatasia (HPP); and
- The following documentation must be included with the request:
    - ALPL molecular genetic testing results
    - Serum alkaline phosphatase (ALP) level
    - Tissue-non-specific alkaline phosphatase (TNSALP) substrate level; AND
- Patient must be clinically diagnosed with perinatal/infantile or juvenile HPP initially prior to 18 years of age; and
- Supporting documentation of diagnosis of perinatal/infantile- or juvenile onset HPP prior to 18 years old must be provided; and
- Patient has clinical signs and/or symptoms of hypophosphatasia as supported by clinical notes provided (see appendix A); and
- Diagnosis is supported by one of the following:
    - Molecular genetic testing supporting the presence of mutation in the ALPL gene detected; or
    - Diagnosis is supported by ALL of the following (provided with submitted request):
        - Radiographic imaging provided that demonstrates skeletal abnormalities supporting diagnosis of hypophophatasia (e.g., infantile rickets, alveolar bone

loss, osteoporosis, low bone mineral content for age [as detected by DEXA]) such as the following clinical features; and

- Craniosynostosis (premature fusion of one or more cranial sutures) with increased intracranial pressure;
- Rachitic chest deformity (costochondral junction enlargement seen in advanced rickets) with associated respiratory compromise;
- Limb deformity with delayed walking or gait abnormality;
- Compromised exercise capacity due to rickets and muscle weakness;
- Low bone mineral density for age with unexplained fractures;
- Alveolar bone loss with premature loss of deciduous (primary) teeth.

  - A low baseline serum alkaline phosphatase (ALP) lab results provided supporting level below the gender- and age-specific reference range of the laboratory performing the test; and
  - Elevated TNSALP substrate level as supported by lab results provided (i.e. serum PLP level, serum or urine PEA level, urinary PPi level); and

- Baseline ophthalmology exam; and
- Baseline renal ultrasound; and
- Member weight within 30 days of request; and
- Dose is than no greater than 2mg/kg three (3) times weekly and appropriate vials must be used for patient;
- MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements

Renewal coverage:

- Supporting documentation provided that Strensiq has been effective in management of HPP and patient is responding to treatment such as:
  - Improvements in weight;
  - Improvement in height velocity;
  - Improvement in ventilator status, respiratory function;
  - Improvement in skeletal manifestations (e.g. bone mineralization, bone formation and remodeling, fractures, deformities);
  - Improvement in motor function, mobility or gait;
- Patient is tolerating therapy with Strensiq; and
- Documented ophthalmology exam once yearly to monitor ectopic calcifications; and
- Documented renal ultrasound once yearly to monitor ectopic calcifications.

Coverage durations:

- Initial coverage: 6 months

- Renewal coverage: 6 months

*** Requests will also be reviewed to National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) if applicable.***

Appendix A:

Examples of Signs and Symptoms of HPP

A. Perinatal/infantile-onset HPP:
   - Generalized hypomineralization with rachitic features, chest deformities and rib fractures
   - Skeletal abnormalities (e.g., short limbs, abnormally shaped chest, soft skull bone)
   - Respiratory problems (e.g., pneumonia)
   - Hypercalcemia
   - Failure to thrive
   - Severe muscular hypotonia and weakness
   - Nephrocalcinosis secondary to hypercalciuria
   - Swallowing problems
   - Seizures

Investigational use: All therapies are considered investigational when used at a dose or for a condition other than those that are recognized as medically accepted indications as defined in any one of the following standard reference compendia: American Hospital Formulary Service Drug information (AHFS-DI), Thomson Micromedex DrugDex, Clinical Pharmacology, Wolters Kluwer Lexi-Drugs, or Peer-reviewed published medical literature indicating that sufficient evidence exists to support use. Neighborhood does not provide coverage for drugs when used for investigational purposes.

Applicable Codes:
Below is a list of billing codes applicable for covered treatment options. The below tables are provided for reference purposes and may not be all inclusive. Requests received with codes from tables below do not guarantee coverage. Requests must meet all criteria provided in the procedure section.

The following HCPCS/CPT codes are:

HCPCS/CPT Code	Description
J3490 (NOC)	Unclassified drugs

References:

1. Strensiq (asfotase alfa) (Prescribing Information). New Haven , CT: Alexion Pharmaceuticals, Inc.; 2018 February
2. Whyte MP, Greenberg CR, Salman NJ, et al. Enzyme-replacement therapy in life-threatening hypophosphatasia. N Engl J Med. 2012;366(10):904-913.
3. Whyte MP, Rockman-Greenberg C, Ozono K, et al. Asfotase alfa treatmentimproves survival for perinatal and infantile hypophosphatasia. J Clin Endocrinol Metab. 2016;101(1):334