Strensiq (asfotase alfa) subcutaneous

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 Criteria:

- 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.; 2021 April.
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