Drug Name: Strensiq (asfotase alfa)
Date: 03-2018
Review Date: 5/19

Drug Name:	Strensiq (asfotase alfa)
Prescriber Restrictions:	Prescriber is endocrinologist or specialist in the treatment of perinatal/infantile or juvenile hypophosphatasia (HPP); and
Required Documentation:	• ALPL molecular genetic testing results • Serum alkaline phosphatase (ALP) level • Tissue-non-specific alkaline phosphatase (TNSALP) substrate level
Initial Coverage Criteria	• 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; and • Diagnosis is supported by one of the following: o Molecular genetic testing supporting the presence of mutation in the ALPL gene detected; or o 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 a) Craniosynostosis (premature fusion of one or more cranial sutures) with increased intracranial pressure; b) Rachitic chest deformity (costochondral junction enlargement seen in advanced rickets) with associated respiratory compromise; c) Limb deformity with delayed walking or gait abnormality; d) Compromised exercise capacity due to rickets and muscle weakness; e) Low bone mineral density for age with unexplained fractures; f) 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.
Renewal Coverage Criteria	• Supporting documentation provided that Strensiq has been effective in management of HPP and patient is responding to treatment such as: o Improvements in weight; o Improvement in height velocity; o Improvement in ventilator status, respiratory function; o Improvement in skeletal manifestations (e.g. bone mineralization, bone formation and remodeling, fractures, deformities); o 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.
Dosing Limitations:	• Dosing and dosing frequency is no greater than 2mg/kg three (3) times weekly. • Appropriate vials must be used for patient.
Coverage Duration:	**Initial**: 6 months **Continuation of therapy**: 6 months

Investigational use: All Multiple sclerosis therapies is considered investigational when used at a dose or for a condition other than those that are recognized as medically accepted indications as defined in one of the above listed resources. Neighborhood does not provide coverage for drugs when used for investigational purses.