Drug Name: Strensiq (asfotase alfa)
Date: 03-2018
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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 (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
	  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
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