BlueCross BlueShield
Federal Employee Program
Blue Cross Blue Shield Association
750 9th St NW, Suite 900
Washington, D.C. 20001
1-800-624-5060
Fax 1-877-378-4727

5.30.015

Section:	Prescription Drugs	**Effective Date:**	April 1, 2025
Subsection:	Endocrine and Metabolic Drugs	**Original Policy Date:**	December 4, 2015
Subject:	Strensiq	**Page:**	1 of 4

Last Review Date: March 7, 2025

Strensiq

Description

Strensiq (asfotase alfa)

Background
Strensiq is used to treat hypophosphatasia (HPP), a rare genetic disorder characterized by the abnormal development of bones and teeth. These abnormalities occur due to defective mineralization, the process by which bones and teeth take up minerals such as calcium and phosphorus. These minerals are required for proper hardness and strength. Hypophosphatasia is caused by mutations in the tissue nonspecific alkaline phosphatase gene. Such mutations lead to low levels of the tissue nonspecific alkaline phosphatase (TNSALP) enzyme. This enzyme is needed for the proper development and health of bones and teeth (1). Strensiq is administered via injection three or six times per week (2).

Regulatory Status
FDA-approved indication: Strensiq is a tissue nonspecific alkaline phosphatase indicated for the treatment of patients with perinatal/infantile- and juvenile-onset hypophosphatasia (2).

Strensiq contains a boxed warning for hypersensitivity reactions, including anaphylaxis. If a severe hypersensitivity reaction occurs, discontinue Strensiq and immediately initiate appropriate medical treatment, including use of epinephrine (2).

Patients with HPP are at increased risk for developing ectopic calcifications of the eye and kidneys. Ophthalmology (eye) examinations and renal ultrasounds are recommended at

baseline and periodically during treatment to monitor for signs and symptoms of ophthalmic and renal ectopic calcifications and for changes in vision or renal function (2).

During clinical trials, anti-drug antibodies have been detected in patients receiving treatment with Strensiq using an electrochemiluminescent (ECL) immunoassay. Antibody positive samples were tested to determine the presence of neutralizing antibodies based on in vitro inhibition of the catalytic activity of Strensiq. Formation of anti-drug antibody resulted in a reduced systemic exposure Strensiq (2).

The safety and effectiveness of Strensiq have been established in pediatric patients. The majority of patients in the clinical trials were pediatric patients 1 day to 16 years of age (2).

Related policies

Policy

This policy statement applies to clinical review performed for pre-service (Prior Approval, Precertification, Advanced Benefit Determination, etc.) and/or post-service claims.

Strensiq may be considered **medically necessary** if the conditions indicated below are met.

Strensiq may be considered **investigational** for all other indications.

Prior-Approval Requirements

Diagnoses

Patient must have **ONE** of the following:

1. Perinatal/infantile-onset hypophosphatasia
2. Juvenile-onset hypophosphatasia

AND ALL of the following:

a. Ophthalmology examination at baseline and periodically throughout treatment
b. Renal ultrasound at baseline and periodically throughout treatment
c. Physician agrees to assess patient's improvement in growth and radiographical findings after one year of therapy and discontinue if **NO** improvement is seen

Prior – Approval *Renewal* Requirements

Diagnoses

Patient must have **ONE** of the following:

1. Perinatal/infantile onset hypophosphatasia
2. Juvenile-onset hypophosphatasia

AND ALL of the following:

a. Ophthalmology examinations are done periodically throughout treatment
b. Renal ultrasound are done periodically throughout treatment
c. Documented improvement is seen in growth and radiographical findings

Policy Guidelines

Pre - PA Allowance

None

Prior – Approval Limit

Duration 12 months

Prior – Approval *Renewal* Limits

Duration 2 years

Rationale

Summary

Strensiq is a tissue nonspecific alkaline phosphatase indicated for the treatment of patients with perinatal/infantile- and juvenile-onset hypophosphatasia. Strensiq has a boxed warning for hypersensitivity reactions. Ophthalmology (eye) examinations and renal ultrasounds are recommended at baseline and periodically during treatment to monitor for signs and symptoms of ophthalmic and renal ectopic calcifications and for changes in vision or renal function. The safety and effectiveness of Strensiq have been established in pediatric patients (2).

Prior authorization is required to ensure the safe, clinically appropriate, and cost-effective use of Strensiq while maintaining optimal therapeutic outcomes.

References

1. Hypophosphatasia. Rare disease information. National Organization for Rare Disorders (NORD) website.
2. Strensiq [package insert]. New Haven, CT: Alexion Pharmaceuticals, Inc.; July 2024.

Policy History

Date	Action
December 2015	Addition to PA
March 2016	Annual review Addition of the requirement of the physician agrees to assess patient's improvement in growth and radiographical findings after one year of therapy and discontinue if no improvement is seen Change of duration from lifetime to 12 months for initiation Addition of renewal section for lifetime duration per SME Policy number change from 5.08.15 to 5.30.15
June 2016	Annual review
September 2016	Annual editorial review and reference update
December 2017	Annual editorial review and reference update
November 2018	Annual editorial review and reference update
December 2019	Annual editorial review. Changed renewal approval duration from lifetime to 2 years
December 2020	Annual review and reference update
September 2021	Annual review
September 2022	Annual review
March 2023	Annual review
March 2024	Annual review
March 2025	Annual editorial review and reference update

Keywords

This policy was approved by the FEP® Pharmacy and Medical Policy Committee on March 7, 2025 and is effective on April 1, 2025.