Strensiq® (asfotase alfa) For injection | 40 mg/mL

A Guide to

Common Prior Authorization (PA) Criteria for STRENSIQ® (asfotase alfa)

For the treatment of perinatal/infantile- and juvenile-onset hypophosphatasia (HPP)[1]

Many commercial and public healthcare insurance plans require prior authorization or precertification for use of STRENSIQ for the treatment of patients with perinatal/infantile- and juvenile-onset HPP.[1] Although requirements vary by plan, there are common criteria that may be used for STRENSIQ. Please verify the current requirements for STRENSIQ for HPP, including whether a PA is required, with each individual plan.

PA Process Tips

Contact your Alexion Field Reimbursement Manager for information about plan-specific PA requirements or general questions about submitting PA requests.

For personalized support on behalf of a specific patient, the patient must be enrolled in OneSource™, Alexion's patient support program, and provide consent for these optional services. Your Field Reimbursement Manager will be able to provide educational support for patient-specific services once the OneSource™ enrollment form is submitted and approved.

INDICATION

STRENSIQ® (asfotase alfa) is indicated for the treatment of patients with perinatal/infantile- and juvenile-onset hypophosphatasia (HPP).

IMPORTANT SAFETY INFORMATION INCLUDING BOXED WARNING

WARNING: HYPERSENSITIVITY REACTIONS INCLUDING ANAPHYLAXIS

Patients treated with enzyme replacement therapies have experienced life-threatening hypersensitivity reactions, including anaphylaxis. Anaphylaxis has occurred during the early course of enzyme replacement therapy and after extended duration of therapy.

Initiate STRENSIQ under the supervision of a healthcare provider with appropriate medical monitoring and support measures. If a severe hypersensitivity reaction (e.g., anaphylaxis) occurs, discontinue STRENSIQ and immediately initiate appropriate medical treatment, including use of epinephrine. Inform patients of the symptoms of life-threatening hypersensitivity reactions, including anaphylaxis and to seek immediate medical care should symptoms occur *[see Warnings and Precautions (5.1)]*.

WARNINGS AND PRECAUTIONS

- **Life-threatening hypersensitivity reactions, including anaphylaxis**, have been reported in STRENSIQ-treated patients. Signs and symptoms consistent with anaphylaxis included difficulty breathing, choking sensation, nausea, periorbital edema, and dizziness. These reactions have occurred within minutes after subcutaneous administration of STRENSIQ and have been observed more than 1 year after treatment initiation. Other hypersensitivity reactions have also been reported in STRENSIQ-treated patients, including vomiting, fever, headache, flushing, irritability, chills, erythema, rash, pruritus, and oral hypoesthesia. Consider the risks and benefits of re-administering STRENSIQ following a severe reaction. If the decision is made to re-administer, monitor patients for a reoccurrence of signs and symptoms of a severe hypersensitivity reaction.

Commercial and Public Healthcare Insurance Plans

Below are common criteria that are required by many healthcare insurance plans including commercial, Federal Employee Program (FEP), Medicaid, Medicare Advantage-Prescription Drug (MA-PD), Medicare Part D, Pharmacy Benefit Manager (PBM), and TRICARE plans.

Date of Onset of Symptoms[1]

- Perinatal/infantile- or juvenile-onset of HPP disease is required. If the patient is 18 years of age or older, provide documentation of the presence of the condition prior to 18 years of age

Relevant Lab Results for HPP[2,3]

Most plans require confirmation of:

- Low serum ALP activity
- Skeletal abnormalities
- Substrate elevation (plasma PLP [also called B_6], and urine PEA)

Some plans may require:

- Genetic testing documenting tissue-nonspecific alkaline phosphatase (*ALPL*) gene mutation
- Radiographic imaging of skeletal abnormalities

Current or History of Clinical Manifestations of HPP[2,3]

Patients may present with or report a history of symptoms that affect one or more areas of the body:

- Generalized hypomineralization with rachitic fractures, chest deformities, and rib fractures
- Skeletal abnormalities
- Respiratory problems
- Hypercalcemia/hypercalciuria
- Failure to thrive
- Hypotonia and weakness
- Nephrocalcinosis
- Poor feeding
- Seizures

Signs and symptoms for infantile- or juvenile-onset patients can include:

- Poor dentition
- Failure to thrive
- Joint, bone, and muscle pain
- Muscle weakness
- Frequent fractures
- Mobility issues

Adult patients with perinatal/infantile- and/or juvenile-onset HPP may report a history of the above symptoms. They may also report additional symptoms or progression of symptoms as an adult.

Patient's Weekly Dose Will Not Exceed[1]

- 9 mg/kg for perinatal/infantile-onset HPP
- 6 mg/kg weekly for juvenile-onset HPP

Who May Prescribe?

Some plans may require that STRENSIQ is prescribed by or in consultation with an endocrinologist, geneticist, or a metabolic disorder specialist. Review the specific policy for any specialist prescribing limitations; these could include prescribing by or in consultation with a metabolic bone and mineral specialist, specialist in HPP or related disorders, and pediatric specialists including orthopedic surgeons, pediatric endocrinologists, neonatologists, and/or medical geneticists.

Additional Information

Medical necessity exceptions may be available even if your patient does not meet all of the criteria.

Continued coverage of STRENSIQ generally requires follow-up with an appropriate specialist and a positive clinical response from baseline for the appropriate indication. **Please always refer to the specific health plan requirements for each patient.**

Our dedicated Field Reimbursement Managers (FRMs) can work with you

If you have questions about the prior authorization process or specific questions about a particular plan, please reach out to your FRM.

ALP = alkaline phosphatase; PEA = phosphoethanolamine; PLP = pyridoxal 5'-phosphate.

Please see Important Safety Information on pages 1 and 4 and full Prescribing Information for STRENSIQ (asfotase alfa), including Boxed WARNING regarding hypersensitivity reactions including anaphylaxis.

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Some Codes That May Be Appropriate for the STRENSIQ® (asfotase alfa) Indication*

ICD-10 diagnosis codes†

E83.31 Familial hypophosphatemia (perinatal/infantile- and juvenile-onset hypophosphatasia [HPP])‡

E83.39 Other disorders of phosphorus metabolism (perinatal/infantile- and juvenile-onset hypophosphatasia [HPP])‡

CPT codes§

76770-76775 Ultrasound, retroperitoneal (eg, renal, aorta, nodes), real time with image documentation; complete; limited

76811 Ultrasound, pregnant uterus, real time with image documentation, fetal and maternal evaluation plus detailed fetal anatomic examination, transabdominal approach; single or first gestation

+76812 Each additional gestation (list separately in addition to code for primary procedure)

77075 Radiologic examination, osseous survey; complete (axial and appendicular skeleton)

81404 81406 81479 Hypophosphatasia and hypophosphatemic rickets panel

84207 Pyridoxal 5'-phosphate (PLP), also known as Vitamin B_6

84075 Alkaline phosphatase (ALP), total

92012-92014 Ophthalmological services: medical examination and evaluation with initiation or continuation of diagnostic and treatment program; intermediate, established patient; 1 or more visits

96372 Therapeutic, prophylactic, or diagnostic injection (specify substance or drug); subcutaneous or intramuscular

STRENSIQ Vial Presentations[1]

Strength	Quantity (Vials per carton)	National Drug Code (NDC)
18 mg/0.45 mL	1	25682-010-01
	12	25682-010-12
28 mg/0.7 mL	1	25682-013-01
	12	25682-013-12
40 mg/mL	1	25682-016-01
	12	25682-016-12
For patients ≥40 kg		
80 mg/0.8 mL	1	25682-019-01
	12	25682-019-12

Important Reminder

In order to facilitate a timely review of the PA request when one is required, be sure to submit all requisite documentation together with the fully completed PA/precertification form.

Providers are responsible for timely and accurate submission of PA requests. Alexion does not make any representation or guarantee concerning reimbursement or coverage for any service or item.

*Source: Information is based on a review of 2024 coverage and PA criteria for national and large regional US commercial, Federal Employee Program (FEP), Medicare Advantage-Prescription Drug (MA-PD), Medicare Part D, Managed Medicaid, Pharmacy Benefit Manager (PBM) plans, and TRICARE. Please check with the individual payer for specific coverage information because coverage policies change, and information can vary.

†These ICD-10 diagnosis codes are not intended to be promotional or to encourage or suggest a use of drug that is inconsistent with FDA-approved use. The codes provided are not exhaustive and additional codes may apply. Descriptions in parentheses were added by Alexion for clarification and do not appear in these ICD-10 codes.

‡Information in parentheses added for clarity.

§CPT = Current Procedural Terminology. CPT® is a registered trademark of the American Medical Association, 2020.

This resource is provided for informational purposes only and is not medical advice or guidance. It is not inclusive of all payer prior authorization or precertification criteria for STRENSIQ for HPP. Alexion does not warrant, promise, guarantee, or make any statement that the use of this information will result in coverage or payment for STRENSIQ, or that any payment received will cover providers' costs.

IMPORTANT SAFETY INFORMATION INCLUDING BOXED WARNING (cont'd)

WARNINGS AND PRECAUTIONS (cont'd)

- **Lipodystrophy:** Localized lipodystrophy, including lipoatrophy and lipohypertrophy has been reported at injection sites after several months in patients treated with STRENSIQ in clinical trials. Advise patients to follow proper injection technique and to rotate injection sites.
- **Ectopic Calcifications:** Patients with HPP are at increased risk for developing ectopic calcifications. Events of ectopic calcification, including ophthalmic (conjunctival and corneal) and renal (nephrocalcinosis, nephrolithiasis), have been reported in the clinical trial experience with STRENSIQ. There was insufficient information to determine whether the reported events were consistent with the disease or due to STRENSIQ. No visual changes or changes in renal function were reported resulting from the occurrence of ectopic calcifications.

  Ophthalmology examinations and renal ultrasounds are recommended at baseline and periodically during treatment with STRENSIQ to monitor for signs and symptoms of ophthalmic and renal ectopic calcifications and for changes in vision or renal function.
- **Possible Immune-Mediated Clinical Effects:** In clinical trials, most STRENSIQ-treated patients developed anti-asfotase alfa antibodies and neutralizing antibodies which resulted in reduced systemic exposure of asfotase alfa. In postmarketing reports, some STRENSIQ-treated patients with initial therapeutic response subsequently developed recurrence and worsening in disease-associated laboratory and radiographic biomarkers (some in association with neutralizing antibodies) suggesting possible immune-mediated effects on STRENSIQ's pharmacologic action resulting in disease progression. The effect of anti-asfotase alfa antibody formation on the long-term efficacy of STRENSIQ is unknown. There are no marketed anti-asfotase alfa antibody tests. If patients experience progression of HPP symptoms or worsening of disease-associated laboratory and imaging biomarkers after a period of initial therapeutic response to STRENSIQ, consider obtaining anti-asfotase alfa antibody testing by contacting STRENSIQ Medical Information at Alexion at 1-888-765-4747 or by email at medinfo@alexion.com. Close clinical follow up is recommended.

ADVERSE REACTIONS

In clinical trials, the most common adverse reactions (≥ 10%) reported were injection site reactions (63%), lipodystrophy (28%), ectopic calcifications (14%), and hypersensitivity reactions (12%). Possible immune-mediated clinical effects have been identified during post-approval use of STRENSIQ.

DRUG INTERACTIONS

Drug Interference with Laboratory Tests:

- Laboratory tests utilizing alkaline phosphatase (ALP) as a detection reagent could result in erroneous test results for patients receiving treatment due to the presence of asfotase alfa in clinical laboratory samples. Inform laboratory personnel that the patient is being treated with STRENSIQ and discuss use of an alternative testing platform which does not utilize an ALP-conjugated test system.
- Elevated serum ALP measurements detected through clinical laboratory testing are expected in patients receiving STRENSIQ due to circulating concentrations of asfotase alfa. Do not rely on serum ALP measurements for clinical decision making in patients treated with STRENSIQ.

SPECIAL POPULATIONS

- **Pregnancy & Lactation:** There are no available data on STRENSIQ use in pregnant women, the presence of STRENSIQ in human milk, or the effects on the breastfed infant or on milk production, to inform a drug associated risk.

To report SUSPECTED ADVERSE REACTIONS, contact Alexion Pharmaceuticals, Inc. at 1-844-259-6783 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch

Please see full Prescribing Information for STRENSIQ (asfotase alfa), including Boxed WARNING regarding hypersensitivity reactions including anaphylaxis.

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References: 1. STRENSIQ. Package insert. Alexion Pharmaceuticals, Inc. **2.** Rockman-Greenberg C. Hypophosphatasia. *Pediatr Endocrinol Rev*. 2013;10(suppl 2):380-388. **3.** Bianchi ML, Bishop NJ, Guañabens N, et al. Hypophosphatasia in adolescents and adults: overview of diagnosis and treatment. *Osteoporos Int*. 2020;31(8):1445-1460.

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