



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

IMPORTANT SAFETY INFORMATION

WARNINGS AND PRECAUTIONS

- **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, skin erythema, rash, pruritus and oral hypoesthesia.

Inform patients and/or caregivers of the signs and symptoms of hypersensitivity reactions and have them seek immediate medical care should signs and symptoms occur. If a severe hypersensitivity reaction occurs, discontinue STRENSIQ treatment and initiate appropriate medical treatment. Consider the risks and benefits of re-administering STRENSIQ to individual patients following a severe reaction. If the decision is made to re-administer the product, monitor patients for a reoccurrence of signs and symptoms of a severe hypersensitivity reaction.

Please see additional Important Safety Information on [page 4](#) and full [Prescribing Information](#) for STRENSIQ (asfotase alfa).

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 Symptoms¹

- 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 B₆], and urine PEA*)

Some plans may require:

- Genetic testing documenting tissue non-specific 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 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 Exceed¹

- 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.

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 | 81404 81406 81479 Hypophosphatasia and hypophosphatemic rickets panel |
| 76811 Ultrasound, pregnant uterus, real time with image documentation, fetal and maternal evaluation plus detailed fetal anatomic examination, transabdominal approach; single or first gestation | 84207 Pyridoxal 5'-phosphate (PLP), also known as Vitamin B ₆ |
| +76812 Each additional gestation (list separately in addition to code for primary procedure) | 84075 Alkaline phosphatase (ALP), total |
| 77075 Radiologic examination, osseous survey; complete (axial and appendicular skeleton) | 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 Presentations¹

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
<i>For patients ≥40 kg</i>		
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 2021 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 parenthesis 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 (cont'd)

WARNINGS AND PRECAUTIONS

- **Lipodystrophy:** Localized lipodystrophy, including lipoatrophy (depression in the skin) and lipohypertrophy (enlargement or thickening of tissue), 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 or not 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

Overall, the most common adverse reactions ($\geq 10\%$) reported were injection site reactions (63%). Other common adverse reactions included 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 and may be unreliable for clinical decision making.

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.

Please see Indication & additional Important Safety Information on [page 1](#) and full [Prescribing Information](#) for STRENSIQ (asfotase alfa).

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References: **1.** STRENSIQ. Package insert. Alexion Pharmaceuticals Inc.; 2020. **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.

