

Clinical Policy: Asfotase Alfa (Strensiq)

Reference Number: LA.PHAR.328

Effective Date: 12.21.23

Last Review Date: 01.15.25

Line of Business: Medicaid

[Coding Implications](#)

[Revision Log](#)

See [Important Reminder](#) at the end of this policy for important regulatory and legal information.

****Please note: This policy is for medical benefit****

Description

Asfotase alfa (Strensiq®) is a tissue nonspecific alkaline phosphatase.

FDA Approved Indication(s)

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

Policy/Criteria

Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.

It is the policy of Louisiana Healthcare Connections that Strensiq is **medically necessary** when the following criteria are met:

I. Initial Approval Criteria

A. Perinatal/Infantile- and Juvenile-Onset Hypophosphatasia (must meet all):

1. Diagnosis of perinatal/infantile- or juvenile-onset HPP as evidenced by all of the following (a, b, and c):
 - a. Age of onset is < 18 years;
 - b. Presence of one of the following laboratory indices (i, ii, or iii):
 - i. Molecular genetic testing documenting tissue non-specific alkaline phosphatase (ALPL) gene mutation;
 - ii. Low baseline serum alkaline phosphatase activity;
 - iii. An elevated level of a tissue non-specific alkaline phosphatase substrate (i.e., serum pyridoxal 5'-phosphate, serum, or urinary inorganic pyrophosphate, urinary phosphoethanolamine);
 - c. History of one of the following HPP clinical manifestations (i, ii, iii, or iv):
 - i. Vitamin B6-dependent seizures;
 - ii. Failure to thrive or growth failure/short stature;
 - iii. Nephrocalcinosis with hypercalcemia/hypercalciuria;
 - iv. Skeletal abnormalities and associated impairments (any of the following):
 - 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;
- 2. Prescribed by or in consultation with an endocrinologist;
 - 3. Dose does not exceed the following (a or b):
 - a. Perinatal/infantile-onset HPP: 9 mg/kg per week;
 - b. Juvenile-onset HPP: 6 mg/kg per week.

Approval duration: 6 months

B. Other diagnoses/indications (must meet 1 or 2):

- 1. If this drug has recently (within the last 6 months) undergone a label change (e.g., newly approved indication, age expansion, new dosing regimen) that is not yet reflected in this policy, refer to LA.PMN.255
- 2. If the requested use (e.g., diagnosis, age, dosing regimen) is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized) AND criterion 1 above does not apply, refer to the off-label use policy for the relevant line of business: LA.PMN.53 for Medicaid.

II. Continued Therapy

A. Perinatal/Infantile- and Juvenile-Onset Hypophosphatasia (must meet all):

- 1. Currently receiving medication via Louisiana Healthcare Connections benefit or member has previously met initial approval criteria;
- 2. Member is responding positively to therapy, as evidenced by improvement in any of the following on initial re-authorization request:
 - a. Height velocity;
 - b. Respiratory function;
 - c. Skeletal manifestations (e.g., bone mineralization, bone formation and remodeling, fractures, deformities);
 - d. Motor function, mobility, or gait;
- 3. If request is for a dose increase, new dose does not exceed the following (a or b):
 - a. Perinatal/infantile-onset HPP: 9 mg/kg per week;
 - b. Juvenile-onset HPP: 6 mg/kg per week.

Approval duration: 12 months

B. Other diagnoses/indications (must meet 1 or 2):

- 1. If this drug has recently (within the last 6 months) undergone a label change (e.g., newly approved indication, age expansion, new dosing regimen) that is not yet reflected in this policy, refer to LA.PMN.255
- 2. If the requested use (e.g., diagnosis, age, dosing regimen) is NOT specifically listed under section III (Diagnoses/Indications for which coverage is NOT authorized) AND criterion 1 above does not apply, refer to the off-label use policy for the relevant line of business: LA.PMN.53 for Medicaid.

III. Diagnoses/Indications for which coverage is NOT authorized:

- A.** Non-FDA approved indications, which are not addressed in this policy, unless there is sufficient documentation of efficacy and safety according to the off label use policy – LA.PMN.53 for Medicaid, or evidence of coverage documents.

IV. Appendices/General Information

Appendix A: Abbreviation/Acronym Key

FDA: Food and Drug Administration

HPP: hypophosphatasia

Appendix B: Therapeutic Alternatives

Not applicable

Appendix C: Contraindications/Boxed Warnings

- Contraindication(s): none reported
- Boxed warning(s): hypersensitivity reactions including anaphylaxis

V. Dosage and Administration

Indication	Dosing Regimen	Maximum Dose
Perinatal/infantile-onset HPP	6 mg/kg SC per week as either: <ul style="list-style-type: none">• 2 mg/kg three times per week, or• 1 mg/kg six times per week <p>The dose may be increased for lack of efficacy (e.g., no improvement in respiratory status, growth, or radiographic findings) up to 9 mg/kg per week, administered as 3 mg/kg SC three times per week.</p>	9 mg/kg/week
Juvenile-onset HPP	6 mg/kg SC per week as either: <ul style="list-style-type: none">• 2 mg/kg three times per week, or• 1 mg/kg six times per week	6 mg/kg/week

VI. Product Availability

Single-use vials: 18 mg/0.45 mL, 28 mg/0.7 mL, 40 mg/mL, 80 mg/0.8 mL

VII. References

1. Strensiq Prescribing Information. New Haven, CT: Alexion Pharmaceuticals, Inc.; July 2024. Available at <http://strensiq-hcp.com>. Accessed July 15, 2024.
2. Beck C, Morback H, Stenzel M. Hypophosphatasia: Recent advances in diagnosis and treatment. *Open Bone J*. 2009; 1:8-15.
3. Scott LJ. Asfotase alfa in perinatal/infantile-onset and juvenile-onset hypophosphatasia: A guide to its use in the USA. *Bio Drugs*. 2016; 30:41-48. DOI 10.1007/s40259-016-0161-x.
4. Whyte MP, Rockman-Greenberg C, Ozono K, et al. Asfotase alfa treatment improves survival for perinatal and infantile hypophosphatasia. *J Clin Endocrinol Metab*. January 2016; 101(1):334-42. Doi: 10.1210/jc.2015-3462. Epub 2015 Nov 3.
5. Orimo H. Pathophysiology of hypophosphatasia and the potential role of asfotase alfa. *Ther Clin Risk Manag*. May 17, 2016; 12:777-86. Doi: 10.2147/TCRM.S87956. eCollection 2016.

6. Mornet E, Nunes ME. Hypophosphatasia. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2016. 2007 Nov 20 [updated 2016 Feb 4]. Available at <https://www.ncbi.nlm.nih.gov/books/NBK1150/>. Accessed August 30, 2017.
7. Bishop N. Clinical management of hypophosphatasia. Clin Cases miner Bone Metab. 2015; 12(2): 170-173.
8. Choida V, Bubbear JS. Update on the management of hypophosphatasia. Ther Adv Musculoskel Dis. 2019;11:1-8.
9. Kishnani PS, et al. Monitoring guidance for patients with hypophosphatasia treated with asfotase alfa. Mol Genetics and Metab. 2017;122:4-17.
10. Khan AA, Brandi ML, Rush ET, et al. Hypophosphatasia diagnosis: current state of the art and proposed diagnostic criteria for children and adults. Osteoporosis International. 2023;35:431-8. <https://doi.org/10.1007/s00198-023-06844-1>.

Coding Implications

Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

HCPCS Codes	Description
J3490, C9399	Unclassified drugs or biologicals

Reviews, Revisions, and Approvals	Date	LDH Approval Date
Converted corporate to local policy.	06.19.23	10.24.23
Annual review: no significant changes; references reviewed and updated.	04.30.24	07.29.24
Annual review: no significant changes; added new Boxed Warning from a recent label update; references reviewed and updated.	01.15.25	

Important Reminder

This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical policy; and other available clinical information. LHCC makes no representations and accepts no liability with respect to the content of any external information used or relied upon in developing this clinical policy. This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved.

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