



Onpattro (patisiran lipid complex) (Intravenous)

Document Number: SHP-0379

Last Review Date: 10/01/2021

Date of Origin: 09/05/2018

Dates Reviewed: 09/2018, 10/2019, 10/2020, 10/2021

I. Length of Authorization

Coverage will be provided for six months and may be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

- Onpattro 10 mg injection: 3 vials every 3 weeks

B. Max Units (per dose and over time) [HCPCS Unit]:

- 300 billable units every 3 weeks

III. Initial Approval Criteria ¹

Coverage is provided in the following conditions:

Submission of medical records (chart notes) related to the medical necessity criteria is **REQUIRED** on all requests for authorizations. Records will be reviewed at the time of submission. Please provide documentation related to diagnosis, step therapy, and clinical markers (i.e. genetic and mutational testing) supporting initiation when applicable. Medical records may be submitted via direct upload through the PA web portal or by fax.

- Patient is at least 18 years of age; **AND**

Universal Criteria ¹

- Patient is receiving supplementation with vitamin A at the recommended daily allowance; **AND**
- Must not be used in combination with other transthyretin (TTR) reducing agents (e.g., inotersen, tafamidis, etc.); **AND**

Polyneuropathy due to Hereditary Transthyretin-Mediated (hATTR) Amyloidosis /Familial Amyloidotic Polyneuropathy (FAP) † Φ ¹⁻⁶

- Patient has a definitive diagnosis of hATTR amyloidosis/FAP as documented by amyloid deposition on tissue biopsy and identification of a pathogenic *TTR* variant using molecular genetic testing; **AND**
- Used for the treatment of polyneuropathy as demonstrated by at least TWO of the following criteria:
 - Subjective patient symptoms are suggestive of neuropathy
 - Abnormal nerve conduction studies are consistent with polyneuropathy
 - Abnormal neurological examination is suggestive of neuropathy; **AND**
- Patient’s peripheral neuropathy is attributed to hATTR/FAP and other causes of neuropathy have been excluded; **AND**
- Baseline in strength/weakness has been documented using an objective clinical measuring tool (e.g., Medical Research Council (MRC) muscle strength, etc.); **AND**
- Patient has not been the recipient of an orthotopic liver transplant (OLT)

† FDA Approved Indication(s); ‡ Compendium Recommended Indication(s) Φ Orphan Drug

IV. Renewal Criteria ¹⁻⁶

Coverage can be renewed based upon the following criteria:

- Patient continues to meet the universal and other indication-specific relevant criteria identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: severe infusion-related reactions, ocular symptoms related to hypovitaminosis A, etc.; **AND**
- Disease response compared to pre-treatment baseline as evidenced by stabilization or improvement in one or more of the following:
 - Signs and symptoms of neuropathy
 - MRC muscle strength

V. Dosage/Administration ¹

Indication	Dose
hATTR/ FAP polyneuropathy	<p><u>Recommended dosage:</u></p> <ul style="list-style-type: none"> • Weight < 100 kg <ul style="list-style-type: none"> ○ 0.3 mg/kg intravenously every 3 weeks • Weight ≥ 100 kg <ul style="list-style-type: none"> • 30 mg intravenously every 3 weeks <p><u>Preparing for Therapy:</u></p> <ul style="list-style-type: none"> • Dosing is based on actual body weight

	<ul style="list-style-type: none"> • Patients should be premedicated with a corticosteroid, acetaminophen and antihistamines. • Infusion should be filtered and diluted and infused, via a pump, over at least 80 minutes.
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VI. Billing Code/Availability Information

HCPCS Code:

- J0222 – Injection, patisiran, 0.1 mg; 1 billable unit = 0.1 mg

NDC:

- Onpattro 10 mg/5 mL single-dose vial: 71336-1000-xx

VII. References

1. Onpattro [package insert]. Cambridge, MA; Alnylam Pharmaceuticals, Inc., May 2021. Accessed August 2021.
2. Adams D, Gonzalez-Duarte A, O’Riordan WD, et al. Patisiran, an RNAi Therapeutic, for Hereditary Transthyretin Amyloidosis. *N Engl J Med*. 2018 Jul 5;379(1):11-21. doi: 10.1056/NEJMoa1716153
3. Adams D, Suhr OB, Dyck PJ, et al. Trial design and rationale for APOLLO, a Phase 3, placebo-controlled study of patisiran in patients with hereditary ATTR amyloidosis with polyneuropathy. *BMC Neurol*. 2017;17(1):181
4. Sekijima Y, Yoshida K, Tokuda T, et al. Familial Transthyretin Amyloidosis. *Gene Reviews*. Adam MP, Ardinger HH, Pagon RA, et al., editors. Seattle (WA): University of Washington, Seattle; 1993-2018.
5. Ando Y, Coelho T, Berk JL, et al. Guideline of transthyretin-related hereditary amyloidosis for clinicians. *Orphanet J Rare Dis*. 2013;8:31.
6. Sekijima Y. Hereditary Transthyretin Amyloidosis. 2001 Nov 5 [updated 2021 Jun 17]. In: Adam MP, Ardinger HH, Pagon RA, Wallace SE, Bean LJH, Mirzaa G, Amemiya A, editors. *GeneReviews®* [Internet]. Seattle (WA): University of Washington, Seattle; 1993–2021.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
E85.1	Neuropathic hereditary familial amyloidosis

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD), Local Coverage Articles (LCAs), and Local Coverage Determinations (LCDs) may exist and compliance with these policies is required where applicable. They can be found at: <http://www.cms.gov/medicare-coverage-database/search/advanced-search.aspx>. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCA/LCD): N/A

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	Applicable State/US Territory	Contractor
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC