Onpattro (patisiran lipid complex)
(Intravenous)

Document Number: IH-0379

Last Review Date: 10/01/2020
Date of Origin: 09/05/2018
Dates Reviewed: 09/2018, 10/2019, 10/2020

I. Length of Authorization

Coverage will be provided initially for nine months and may be renewed for 12 months thereafter.

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:

- Patient must be at least 18 years old; **AND**

Universal Criteria

- 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 [1-5]

- Patient has a definitive diagnosis of hATTR amyloidosis/FAP as documented by 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); **AND**
- Patient is receiving supplementation with vitamin A at the recommended daily allowance

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

IV. Renewal Criteria [1-5]

Authorizations can be renewed based on 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 the following: 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

Indication	Dose

hATTR/ FAP	<u>Recommended dosage:</u>
	• Weight < 100 kg
	  ○ 0.3 mg/kg intravenously every 3 weeks
	• Weight ≥ 100 kg
	  • 30 mg intravenously every 3 weeks
	<u>Preparing for Therapy:</u>
	• Dosing is based on actual body weight
	• 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.
	• Patients should receive vitamin A supplementation.

VI. Billing Code/Availability Information

<u>HCPCS code:</u>

- 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., February 2020. Accessed September 2020.
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.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
E85.1	Neuropathic heredofamilial 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.

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
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