MEDICAL MUTUAL®

Drug Policy

Policy:	**201831-MRx (10/2021)**	**Initial Effective Date: 09/20/2018**
Code(s):	**HCPCS J0222**	**Annual Review Date: 02/20/2024**
SUBJECT:	**Onpattro ® (Patisiran)**	**Last Revised Date: 02/20/2024**

☒Subject to Site of Care

Prior approval is required for some or all procedure codes listed in this Corporate Drug Policy.

Initial and renewal requests for the medication(s) listed in this policy are subject to site of care management. When billed under the medical benefit, administration of the medication will be restricted to a non-hospital facility-based location (i.e., home infusion provider, provider's office, free-standing ambulatory infusion center) unless the member meets the site of care exception criteria. To view the exception criteria and a list of medications subject to site of care management please click here.

OVERVIEW

Onpattro is a lipid nanoparticle formulated RNA interference (RNAi) therapeutic indicated for treatment of hereditary amyloid transthyretin (hATTR) amyloidosis with polyneuropathy. hATTR amyloidosis is a rare, inherited, rapidly-progressive, debilitating, life-threatening disease. It is a multisystem condition caused by mutation in the transthyretin (TTR) gene that results in misfolded TTR protein accumulation (as amyloid) in the nerves, heart, and other areas of the body. Onpattro targets hepatic production of mutant TTR. By reducing the unstable circulating TTR tetramers, organ deposition of amyloid is prevented, thus, stabilizing or improving symptoms of neuropathy.

POLICY STATEMENT

This policy involves the use of Onpattro. Prior authorization is recommended for medical benefit coverage of Onpattro. Approval is recommended for those who meet the conditions of coverage in the **Initial Approval and Renewal Criteria, Preferred Drug (when applicable), Dosing/Administration, Length of Authorization, and Site of Care (when applicable)** for the diagnosis provided. The requirement that the patient meet the Criteria and Preferred Drug for coverage of the requested medication applies to the initial authorization only. All approvals for initial therapy are provided for the initial approval duration noted below; if reauthorization is allowed, a response to therapy is required for continuation of therapy.

RECOMMENDED AUTHORIZATION CRITERIA

Coverage of Onpattro is recommended in those who meet the following criteria:

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:

- 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 or stabilizing agents (e.g., inotersen, tafamidis, vutrisiran, 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 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	**Recommended dosage:**
	• Weight < 100 kg
	  ○ 0.3 mg/kg intravenously every 3 weeks
	• Weight ≥ 100 kg
	  • 30 mg intravenously every 3 weeks

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.

Documentation Requirements:

The Company reserves the right to request additional documentation as part of its coverage determination process. The Company may deny reimbursement when it has determined that the drug provided or services performed were not medically necessary, investigational or experimental, not within the scope of benefits afforded to the member and/or a pattern of billing or other practice has been found to be either inappropriate or excessive. Additional documentation supporting medical necessity for the services provided must be made available upon request to the Company. Documentation requested may include patient records, test results and/or credentials of the provider ordering or performing a service. The Company also reserves the right to modify, revise, change, apply and interpret this policy at its sole discretion, and the exercise of this discretion shall be final and binding.

FOR MEDICAL BENEFIT COVERAGE REQUESTS:

Prior approval is required for HCPCS Codes C9036

Edits and Denials:

Prior approval: Prior approval is required for Onpattro **(HCPCS Code J0222)**. Requests for prior approval will be authorized by a nurse reviewer if submitted documentation meets criteria outlined within the Corporate Medical Policy.

Requests for prior approval will be forwarded to a qualified physician reviewer if submitted documentation does not meet criteria outlined within Corporate Medical Policy.

TOPPS: Claims received with **HCPCS Code J0222** will pend with **Remark Code M3M or M4M** and will be adjudicated in accordance with the Corporate Medical Policy.

Liability: A participating provider will be required to write off charges denied as not medically necessary.

HCPCS Code(s):	
C9036	Injection, patisiran, 0.1 mg (effective 1/1/2019)
J0222	Injection, patisiran, 0.1 mg (effective 10/1/2019)