Policy Title:	Onpattro (patisiran lipid complex) (Intravenous)		
		Department:	PHA
Effective Date:	04/10/2019		
Review Date:	11/27/2019, 1/29/20, 6/10/2021		
Revision Date:	11/27/2019, 1/29/20, 6/10/2021		

Purpose: To support safe, effective and appropriate use of Onpattro (patisiran lipid complex).

Scope: Medicaid, Commercial, Medicare-Medicaid Plan (MMP)

Policy Statement:

Onpattro (patisiran lipid complex) is covered under the Medical Benefit when used within the following guidelines. Use outside of these guidelines may result in non-payment unless approved under an exception process.

Procedure:

Coverage of Onpattro (patisiran lipid complex) will be reviewed prospectively via the prior authorization process based on criteria below.

Initial Criteria:

Polyneuropathy due to Hereditary Transthyretin-Mediated (hATTR) Amyloidosis/Familial Amyloidotic Polyneuropathy (FAP)

- Patient must be at least 18 years old; AND
- Must be prescribed by or in consultation with a neurologist, or physician specializing in the treatment of amyloidosis related to hATTR/FAP; AND
- The patient has both of the following:
  - Diagnosis of hATTR amyloidosis with polyneuropathy
  - Documentation that the patient has a pathogenic TTR mutation (e.g., V30M); AND
- Patient has documentation of one of the following:
  - Patient has a baseline polyneuropathy disability (PND) score ≤ IIIb;
  - Patient has a baseline FAP Stage 1 or 2; AND
- Patient exhibits clinical manifestations of ATTR-FAP (e.g., progressive peripheral sensory-motor polyneuropathy, autonomic neuropathy, motor disability, etc.); 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
- The requested medication will not be used in combination with other transthyretin (TRR) reducing agents [e.g. inotersen (Tegsedi), tafamidis (Vyndaqel/Vyndamax), etc.]; AND
- Dosing is in accordance with FDA prescribing information and does not exceed 0.3mg/kg (30mg maximum) every 3 weeks; AND
- Patient is receiving supplementation with vitamin A at the recommended daily allowance; AND
- Onpattro (patisiran) is unproven and not medically necessary for the treatment of:
    - Sensorimotor or autonomic neuropathy not related to hATTR amyloidosis
    - Primary or leptomeningeal amyloidosis
- MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements

Continuation of Therapy Criteria:

- Meet all initial approval criteria AND is tolerating treatment; 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
- The patient must have demonstrated a beneficial response to treatment with Onpattro therapy compared to baseline [e.g., improvement of neuropathy severity and rate of disease progression as demonstrated by the modified Neuropathy Impairment Scale+7 (mNIS+7) composite score, the Norfolk Quality of Life-Diabetic Neuropathy (QoL-DN) total score, polyneuropathy disability (PND) score, FAP disease stage, manual grip strength]. Documentation from the medical record must be provided.

Coverage durations:

- Initial coverage: 6 months
- Continuation of therapy coverage: 6 months

*** Requests will also be reviewed to National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) if applicable.***

Dosage/Administration:

Indication	**Dose**	**Maximum dose (1 billable unit = 0.1 mg)**
hATTR/ FAP	• Weight < 100 kg: 0.3 mg/kg intravenously every 3 weeks • Weight ≥ 100 kg: 30 mg intravenously every 3 weeks	300 billable units (30 mg) every 3 weeks

Investigational use: All therapies are considered investigational when used at a dose or for a condition other than those that are recognized as medically accepted indications as defined in any one of the following standard reference compendia: American Hospital Formulary Service Drug information (AHFS-DI), Thomson Micromedex DrugDex, Clinical Pharmacology, Wolters Kluwer Lexi-Drugs, or Peer-reviewed published medical literature indicating that sufficient evidence exists to support use. Neighborhood does not provide coverage for drugs when used for investigational purposes.

Applicable Codes:
Below is a list of billing codes applicable for covered treatment options. The below tables are provided for reference purposes and may not be all-inclusive. Requests received with codes from tables below do not guarantee coverage. Requests must meet all criteria provided in the procedure section.

The following HCPCS/CPT codes are:

HCPCS/CPT Code	Description
J0222	Injection, patisiran, 0.1mg

References:

1. Onpattro [package insert]. Cambridge, MA; Alnylam Pharmaceuticals, Inc., May 2021. Accessed June 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.