

# Reimbursement Guide for ONPATTRO<sup>®</sup> (patisiran)



The information is provided as a guide to support payer interaction and reimbursement; however, the level of information required will vary based on key areas that the payer requires be addressed to demonstrate medical necessity.

## Indication

ONPATTRO<sup>®</sup> (patisiran) is indicated for the treatment of the polyneuropathy of hereditary transthyretin-mediated amyloidosis in adults.

## Important Safety Information

### Infusion-Related Reactions

Monitor for signs and symptoms during infusion. Slow or interrupt the infusion if clinically indicated. Discontinue the infusion if a serious or life-threatening infusion-related reaction occurs.

Please see Important Safety Information on the last page and full [Prescribing Information](#).

onpattro<sup>®</sup>  
(patisiran) lipid complex injection  
10 mg/5 mL

# Authorization and Coding Information

Medical chart documentation should be based on each patient's individual history, prior testing results, clinical condition, and actions actually performed by the clinician and other parties.

## Examples of Authorization Documentation and Coverage Parameters





Identify specific documentation that must be submitted with the request	<ul style="list-style-type: none"><li>• Letter of medical necessity</li><li>• Chart notes</li><li>• Specific payer preauthorization/prior authorization form</li><li>• ONPATTRO® (patisiran) Prescribing Information</li><li>• Relevant literature, including published standards of care</li><li>• Clinical documentation related to the disease, which may include:<ul style="list-style-type: none"><li>– Rationale for treatment</li><li>– Summary of patient's medical history<ul style="list-style-type: none"><li>▪ Diagnosis of hereditary transthyretin-mediated (hATTR) amyloidosis with polyneuropathy<ul style="list-style-type: none"><li>▸ Documented transthyretin (TTR) mutation, if known</li><li>▸ Family history of hATTR amyloidosis, if known</li></ul></li><li>▪ Clinical presentation and duration of symptoms<ul style="list-style-type: none"><li>▸ Peripheral sensory-motor neuropathy (describe patient's symptomatology, as appropriate: e.g., tingling or increased pain in the hands/feet, loss of feeling in the hands/feet, numbness or tingling in the wrists, carpal tunnel syndrome, loss of ability to sense temperature, difficulty with fine motor skills, weakness in the legs, difficulty walking)</li><li>▸ Autonomic symptoms (describe patient's symptomatology, as appropriate: e.g., orthostasis, abnormal sweating, sexual dysfunction, recurrent urinary tract infections, dysautonomia [constipation and/or diarrhea, nausea, vomiting, anorexia, early satiety])</li></ul></li></ul></li><li>▪ Baseline ambulatory status (see baseline assessments on following page)</li><li>▪ Current supportive care management</li><li>▪ Other relevant aspects of patient history, including imaging, studies, or assessments</li></ul></li></ul>
Determine the preauthorization/prior authorization coverage parameters	<ul style="list-style-type: none"><li>• Time limits of authorization</li><li>• Diagnosis limitations</li><li>• Submission requirements</li></ul>

# Ambulatory Status Assessments for hATTR Amyloidosis

When documenting a patient's ambulatory status, consider the following assessments. As a reminder, assessment of each patient's clinical condition should be based on the treating physician's professional judgment.






## Familial Amyloid Polyneuropathy (FAP) Stage

Clinical staging system as described by Coutinho et al.,<sup>1</sup> according to sensory and motor neuropathy progression.

Stage	Description
 0	<ul style="list-style-type: none"><li>• No symptoms of sensory or motor neuropathy</li></ul>
 I	<ul style="list-style-type: none"><li>• Unimpaired ambulation; mostly mild sensory, motor, and autonomic neuropathy in the lower limbs</li></ul>
 II	<ul style="list-style-type: none"><li>• Assistance with ambulation required; mostly moderate impairment progression to the lower limbs, upper limbs, and trunk</li></ul>
 III	<ul style="list-style-type: none"><li>• Wheelchair-bound or bedridden; severe sensory, motor, and autonomic involvement of all limbs</li></ul>

## Polyneuropathy Disability (PND) Score

Modified PND scoring system first described by Yamamoto et al.<sup>2</sup> to assess the neuropathy of patients with hATTR amyloidosis.

Stage	Description
 0	<ul style="list-style-type: none"><li>• No symptoms of neuropathy</li></ul>
 I	<ul style="list-style-type: none"><li>• Sensory disturbances but preserved walking capability</li></ul>
 II	<ul style="list-style-type: none"><li>• Impaired walking capacity but ability to walk without a stick or crutches</li></ul>
 III	<ul style="list-style-type: none"><li>• A: Walking with the help of one stick or crutch</li><li>• B: Walking with the help of two sticks or crutches</li></ul>
 IV	<ul style="list-style-type: none"><li>• Confined to a wheelchair or bedridden</li></ul>

## Coding Information

Please refer to the information below to support appropriate claims submission for ONPATTRO® (patisiran).

### ONPATTRO

NDC: 71336-1000-01

HCPCS: J3490 Unclassified drugs

C9036 Injection, patisiran, 0.1 mg

### Procedure Codes (select as appropriate):

- 96365 IV infusion, for therapy, prophylaxis, or diagnosis; initial, up to 1 hour
- +96366 IV infusion, for therapy, prophylaxis, or diagnosis; each additional hour
- +96367 IV infusion, for therapy, prophylaxis, or diagnosis; additional sequential infusion of a new drug/substance; up to 1 hour (for premedications)
- 96379 Unlisted therapeutic, prophylactic, or diagnostic intravenous or intra-arterial injection or infusion
- 96413 Chemotherapy administration, IV infusion technique; up to 1 hour, single or initial substance/drug
- +96415 Chemotherapy administration, IV infusion technique; each additional hour

### Additional Information When Billing

- Billing UOM for unclassified or unlisted drugs (J3490) = '1'
- Diagnosis (ICD-10 Codes):
  - E85.1, Neuropathic hereditary amyloidosis
- Dosage UOM = 'mL'; Strength UOM = 'mg'
  - ONPATTRO is administered via intravenous (IV) infusion once every 3 weeks. For patients weighing <100 kg, the recommended dose is 0.3 mg/kg. For patients weighing ≥100 kg, the recommended dose is 30 mg

# Dose Calculation

How to calculate mg	How to calculate mL
$(\text{body weight [kg]} \times 0.3 \text{ mg/kg}) = \text{mg}$	$(\text{mg} \times 5 \text{ mL}/10 \text{ mg}) = \text{mL}$

Proper preparation of ONPATTRO® (patisiran) requires filtration to remove particulates. An additional vial may be required depending on the type of filter used and the amount of product that remains in the filter (hold-up volume). The calculator found in the ONPATTRO Dosing & Administration Guide assumes that 1 mL of drug product remains in the filter when determining the number of vials needed.

## Example - 68 kg Patient

mg	mL
$68 \text{ kg} \times 0.3 \text{ mg/kg} = 20.4 \text{ mg}$	$20.4 \text{ mg} \times 5 \text{ mL}/10 \text{ mg} = 10.2 \text{ mL}$

Some payers may require the use of a JW modifier when billing for the unused portion of the single-dose vial (wastage)—providers should contact payers about specific coding and payment policies.

Please refer to the **ONPATTRO Billing and Coding Guide—Physician Office** or the **ONPATTRO Billing and Coding Guide—Hospital Outpatient Department** for a detailed overview of billing guidelines.

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## Important Safety Information

### Infusion-Related Reactions

Infusion-related reactions (IRRs) have been observed in patients treated with ONPATTRO. In a controlled clinical study, 19% of ONPATTRO-treated patients experienced IRRs, compared to 9% of placebo-treated patients. The most common symptoms of IRRs with ONPATTRO were flushing, back pain, nausea, abdominal pain, dyspnea, and headache.

To reduce the risk of IRRs, patients should receive premedication with a corticosteroid, acetaminophen, and antihistamines (H1 and H2 blockers) at least 60 minutes prior to ONPATTRO infusion. Monitor patients during the infusion for signs and symptoms of IRRs. If an IRR occurs, consider slowing or interrupting the infusion and instituting medical management as clinically indicated. If the infusion is interrupted, consider resuming at a slower infusion rate only if symptoms have resolved. In the case of a serious or life-threatening IRR, the infusion should be discontinued and not resumed.

### Reduced Serum Vitamin A Levels and Recommended Supplementation

ONPATTRO treatment leads to a decrease in serum vitamin A levels. Supplementation at the recommended daily allowance (RDA) of vitamin A is advised for patients taking ONPATTRO. Higher doses than the RDA should not be given to try to achieve normal serum vitamin A levels during treatment with ONPATTRO, as serum levels do not reflect the total vitamin A in the body.

Patients should be referred to an ophthalmologist if they develop ocular symptoms suggestive of vitamin A deficiency (e.g. night blindness).

### Adverse Reactions

The most common adverse reactions that occurred in patients treated with ONPATTRO were upper respiratory tract infections (29%) and infusion-related reactions (19%).

For additional information about ONPATTRO, please see the full [Prescribing Information](#).

**References:** 1. Coutinho P, Martins da Silva A, Lopes Lima JL, et al. In: Glenner G, Pinho e Costa P, Falcão de Freitas A, eds. *Amyloid and amyloidosis: Proceedings of the Third International Symposium on Amyloidosis*. Póvoa de Varzim, Portugal, 23-28 September 1979. Amsterdam: Excerpta Medica. 1980:88-98. 2. Yamamoto S, Wilczek HE, Nowak G, et al. *Am J Transplant*. 2007;7(11):2597-2604.