Amyloidosis Therapy
Onpattro (patisiran)
Tegsedi (inotersen)
Effective 02/01/2023

Plan	☐ MassHealth ☒ MH UPPL ☐Commercial/Exchange	**Program Type**	☒ Prior Authorization ☐ Quantity Limit ☐ Step Therapy
Benefit	☒ Pharmacy Benefit ☒ Medical Benefit (NLX)		
Specialty Limitations	N/A		
Contact Information	**Specialty Medications**		
	All Plans	Phone: 866-814-5506	Fax: 866-249-6155
	Non-Specialty Medications		
	MassHealth	Phone: 877-433-7643	Fax: 866-255-7569
	Commercial	Phone: 800-294-5979	Fax: 888-836-0730
	Exchange	Phone: 855-582-2022	Fax: 855-245-2134
	Medical Specialty Medications (NLX)		
	All Plans	Phone: 844-345-2803	Fax: 844-851-0882
Exceptions	Onpattro injection is available through medical benefit.		

Overview

Onpattro® (patisiran) contains a transthyretin-directed small interfering RNA (siRNA) and is indicated for the treatment of polyneuropathy of hereditary transthyretin-mediated (hATTR) amyloidosis in adults.

Tegsedi® (inotersen) is a transthyretin-directed antisense oligonucleotide indicated for treatment of the polyneuropathy of hATTR amyloidosis in adults.

No PA	Drugs that require PA
	Onpattro® (patisiran)[PD] Tegsedi® (inotersen)

[PD] Preferred Drug. In general, a trial of the preferred drug or clinical rationale for prescribing a non-preferred drug within a therapeutic class.

Coverage Guidelines

Authorization may be reviewed on a case by case basis for members new to the plan who are currently receiving treatment with the requested medication excluding when the product is obtained as samples or via manufacturer's patient assistance programs.

OR

Authorization may be granted for members when ALL the following criteria are met, and documentation is provided:

Onpattro® (patisiran)

1. Diagnosis of hATTR amyloidosis
2. Member is ≥18 years of age
3. Member's current weight (used to verify correct dosing)
4. Documentation of baseline polyneuropathy disability (PND) score of I, II, IIIa, or IIIb†
5. Appropriate dosing

Tegsedi® (inotersen)

1. Diagnosis of hATTR amyloidosis
2. Member is ≥18 years of age
3. Prescriber is a rheumatologist or neurologist, or specialist consult notes from a rheumatologist or neurologist are provided
4. Results from genetic testing showing mutations in the TTR gene
5. Physician documentation of inadequate response, adverse reaction, or contraindication to **ONE** or contraindication to **BOTH** of the following
    a. Amvuttra® (vutrisiran)
    b. Onpattro® (patisiran)
6. Documentation of baseline polyneuropathy disability (PND) score of I, II, IIIa, or IIIb†
7. Appropriate dosing

Continuation of Therapy

Reauthorizations will be granted with documentation of **ALL** of the following:

1. Documentation of positive response to therapy
2. For Onpattro: Updated member weight
3. If member is stable on Tegsedi, the member must meet initial approval criteria listed above.

† The polyneuropathy disability score is an additional assessment tool with ranking based on classes I-IV. Higher scores are indicative of more impaired walking ability. The classes are defined as follows:

I: preserved walking, sensory disturbances
II: impaired walking without need for a stick or crutches
IIIa: walking with one stick or crutch
IIIb: walking with two sticks or crutches
IV: confined to wheelchair or bedridden

Limitations

1. Initial approvals and reauthorizations for:
    a. Onpatrro: 12 months
    b. Tegsedi: 6 months

Appendix

Appendix A: Dosing for Onpattro

Onpattro 10mg/5mL	IV: Dosing is based on actual body weight
	< 100kg: 0.3mg/kg once every 3 weeks
	≥ 100kg: 30mg once every 3 weeks
Tegsedi 284mg/1.5mL	284mg once weekly

References

1. Onpattro® [package insert] San Diego (CA): Alnylam Pharmaceuticals; 2021 May.

2. Tegsedi® [package insert] Boston (MA): Akcea Therapeutics, Inc; 2020 Sep.
3. Hawkins PN, Ando Y, Dispenzeri A, et al. Evolving landscape in the management of transthyretin amyloidosis. Ann Med. 2015;47(8):625-38.
4. Plante-Bordeneuve V. Update in the diagnosis and management of transthyretin familial amyloid polyneuropathy. J Neurol. 2014 Jun;261(6):1227-33.
5. Benson M. Liver transplantation and transthyretin amyloidosis. Muscle Nerve. 2013. 47:157–162.
6. Adams D, Suhr OB, Hund E, et al. First European consensus for diagnosis, management, and treatment of transthyretin familial amyloid polyneuropathy. Current opinion in neurology. 2016;29 Suppl 1:S14-26.
7. Ando Y, Coelho T, Berk JL, et al. Guideline of transthyretin-related hereditary amyloidosis for clinicians. Orphanet J Rare Dis. 2013;8:31.
8. Siddiqi OK, Ruberg FL. Cardiac amyloidosis: an update on pathophysiology, diagnosis, and treatment. Trends Cardiovasc Med. 2018; 28(1):10-21.
9. Gonzalez-Lopez E, Lopez-Sainz A, Garcia-Pavia P. Diagnosis and treatment of transthyretin cardiac amyloidosis. Rev Esp Cardiol. 2017; 70(11):991-1004.
10. Ando Y, Coelho T, Berk JL, et al. Guideline of transthyretin-related hereditary amyloidosis for clinicians. Orphanet J Rare Dis. 2013; 8:31.
11. Nativi-Nicolau J, Maurer MS. Amyloidosis cardiomyopathy: update in the diagnosis and treatment of the most common types. Curr Opin Cardiol. 2018; 33(5): 571-579.
12. Brunjes DL, Castano A, Clemons A, et al. Transthyretin cardiac amyloidosis in older Americans. J Card Fail. 2016; 22(12): 996-1003.
13. Ruberg FL, Maurer MS, Judge DP, et al. Prospective evaluation of the morbidity and mortality of wilf-type and V122I mutant transthyretin amyloid cardiomyopathy: the Transthyretin Amyloidosis Cardiac Study (TRACS). Am Heart J. 2012; 164(2): 222-228.
14. Fontana M. Cardiac amyloidosis: Clinical manifestations and diagnosis. In: Basow D (Ed). UpToDate [database on the Internet]. Waltham (MA): UpToDate: 2020 [cited 2021 Aug 26]. Available from: http://www.utdol.com/utd/index.do.
15. Hafeez AS, Bavry AA. Diagnosis of Transthyretin Amyloid Cardiomyopathy. Cardiol Thep. 2020 Jun;9(1):85- 85.

Review History

05/19/2021 – Created and Reviewed; separated out MH vs. Comm/Exch criteria. Effective 07/01/2021.
11/17/2021 – Reviewed and updated; added Tegsedi to policy. Matched MH UPPL effective 1/1/2022.
11/16/2022 – Reviewed and updated for Nov P&T. Matched MH UPPL. Criteria for Tegsedi updated to require a trial with either Onpattro or Amvuttra. Member stable on Tegsedi must meet initial criteria. Updated references. Effective 2/1/2023.

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