JOHNS HOPKINS MEDICINE / JOHNS HOPKINS HEALTHCARE	Johns Hopkins HealthCare LLC **Pharmacy Public** **Pharmacy Management Drug Policies**	*Policy Number*	MEDS116
		Effective Date	07/17/2019
		Review Date	07/17/2019
	Subject **Tegsedi**	*Revision Date*	12/08/2021
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This document applies to the following Participating Organizations:

Priority Partners

Keywords: Tegsedi

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I. POLICY

Tegsedi (Inotersen) will require prior authorization to ensure appropriate use. The process for initiating a prior authorization request can be found in policy PHARM 20.

1. PPMCO members are subject to the Priority Partners formulary, available at www.ppmco.org.
2. USFHP members are subject to prior authorization criteria, step-edits and days-supply limits outlined in the Tricare Policy Manual. Tricare Policy supersedes JHHC Medical/Pharmacy Policies. Tricare limits may be accessed at: http://pec.ha.osd.mil/formulary_search.php?submenuheader=1

II. POLICY CRITERIA

A. Tegsedi may be approved for patients meeting the following:
   1. Patient is 18 years of age or older
   2. Diagnosis of hereditary transthyretin-mediated amyloidosis with polyneuropathy
   3. Confirmation of transthyretin (TTR) mutation through genetic testing
   4. Documented clinical signs and symptoms of the disease(such as motor disability, peripheral polyneuropathy, autonomic neuropathy)
   5. Documentation of one of the following:
      a. Patient has a baseline polyneuropathy disability (PND) score less than or equal to IIIb
      b. Patient has a baseline familial amyloidotic polyneuropathy(FAP) of Stage 1 or 2
      c. Patient has a baseline neuropathy impairment score (NIS) between 10 and 130
   6. Documented baseline platelet count greater than or equal to 100 x 10^9/L
   7. Documented urinary protein to creatinine ratio (UPCR) of 1000 mg/g or higher
   8. Prescriber is, or has consulted with a neurologist

III. AUTHORIZATION PERIOD/LIMITATIONS

A. Initial approval will be restricted to 6 months of therapy.
B. Authorization for a continuation of therapy can be approved in 12-month intervals with documentation showing clinical benefit from treatment. Clinical benefit is evidenced by improved neurologic impairment, motor function, quality of life, or slowing of disease progression, as well as improvement or stabilization of one of the following:
   1. Polyneuropathy disability (PND) score less than or equal to IIIb

2. Familial amyloidotic polyneuropathy(FAP) of Stage 1 or 2
3. Neuropathy impairment score (NIS) between 10 and 130

IV. EXCLUSIONS

A. Tegsedi will NOT be approved for patients with the following:
   1. History of acute glomerulonephritis caused by Tegsedi
   2. Platelet count less than 100 x 10^9/L
   3. Concurrent use with Tafamidis meglumine, or Oligonucleotide agents, such as Onpattro (patisiran)
   4. History of liver transplant
   5. Severe renal impairment or ESRD
   6. Moderate to severe hepatic impairment
   7. Sensorimotor or autonomic neuropathy not related to hATTR amyloidosis

B. The use of physician samples, or manufacturer product discounts, does not guarantee coverage under the provisions of the medical and/or pharmacy benefit. All pertinent criteria must be met in order to be eligible for benefit coverage.

V. REFERENCES

1. Tegsedi [prescribing information]. Ionis Pharmaceuticals, Inc: Carlsbad, CA; October 2018.
2. Benson MD, Waddington-Cruz M, Berk JL, et al. Inotersen Treatment for Patients with Hereditary Transthyretin Amyloidosis. N Engl J Med. 2018 Jul 5;379(1):22-31.
3. Institute for Clinical and Economic Review. Inotersen and Patisiran for Hereditary Transthyretin Amyloidosis: Effectiveness and Value Final Evidence Report. lPublished October 2018. Available at: https://icer-review.org/wp-content/uploads/2018/02/ICER_Amyloidosis_Final_Evidence_Report_100418.pdf
4. Ando Y, Coelho T, Berk JL, et al. Guideline of transthyretin-related hereditary amyloidosis for clinicians. Orphanet J Rare Dis. 2013;8:31. Available at:https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3584981/

VI. APPROVALS

Signature on file at JHHC

DATE OF REVISION	SUMMARY OF CHANGE
07/17/2019	Policy Creation
12/08/2021	Updated Exclusions section regarding physician samples

Review Date: 07/17/2019

Revision Date: 07/17/2019, 12/08/2021