BlueCross BlueShield Federal Employee Program

TEGSEDI (inotersen)

Pre - PA Allowance

None

Prior-Approval Requirements

Age 18 years of age or older

Diagnosis

Patient must have the following:

Polyneuropathy of hereditary transthyretin-mediated (hATTR) amyloidosis

AND ALL of the following:

1. Diagnosis of hATTR confirmed by genetic testing **OR** tissue biopsy showing amyloid deposition
2. Patient must have **ONE** of the following baseline scores:
    a. Polyneuropathy disability (PND) score ≤ IIIb (see Appendix 1)
    b. FAP Stage 1 or 2 (see Appendix 2)
3. Platelet count ≥ 100 x 10^9/L (100,000 cells/µL)
4. eGFR ≥ 45 mL/minute/1.73 m^2
5. Prescriber agrees to monitor the following during therapy:
    a. Platelet count
    b. Renal function (serum creatinine, eGFR, and urinalysis)
    c. Liver function (ALT, AST, and total bilirubin)
6. Patient and prescriber are both enrolled in the Tegsedi REMS Program
7. Prescriber agrees to supplement the patient with the recommended daily allowance of Vitamin A if indicated
8. Patient has **NONE** of the following:
    a. New York Heart Association (NYHA) class III or IV heart failure
    b. Sensorimotor or autonomic neuropathy not related to hATTR amyloidosis (monoclonal gammopathy, autoimmune disease, etc.)
    c. Prior liver transplantation
9. Prescribed by or in consultation with a neurologist, or a specialist in the treatment of the patient's diagnosis
10. **NO** dual therapy with another Prior Authorization (PA) medication for polyneuropathy caused by hATTR amyloidosis (see Appendix 3)

Prior - Approval Limits

Quantity 12 prefilled syringes per 84 days

Duration 12 months

Prior – Approval *Renewal* Requirements

Age 18 years of age and older

Diagnosis

Patient must have the following:

Polyneuropathy of hereditary transthyretin-mediated (hATTR) amyloidosis

AND ALL of the following:

1. Patient condition has improved or stabilized
2. Platelet count ≥ 100 x 10^9/L (100,000 cells/µL)
3. eGFR ≥ 45 mL/minute/1.73 m^2
4. Prescriber agrees to monitor the following during therapy:
    a. Platelet count
    b. Renal function (serum creatinine, eGFR, and urinalysis)
    c. Liver function (ALT, AST, and total bilirubin)
5. Patient and prescriber are both enrolled in the Tegsedi REMS Program
6. Prescriber agrees to supplement the patient with the recommended daily allowance of Vitamin A if indicated
7. **NO** dual therapy with another Prior Authorization (PA) medication for polyneuropathy caused by hATTR amyloidosis (see Appendix 3)

Prior - Approval *Renewal* Limits

Same as above

Appendix 1 - Polyneuropathy Disability (PND) Severity Scoring System

Polyneuropathy Disability (PND) Score	
Stage 0	No impairment
Stage I	Sensory disturbances but preserved walking capability
Stage II	Impaired walking capability but ability to walk without a stick or crutches
Stage IIIA	Walking only with the help of one stick or crutches
Stage IIIB	Walking only with the help of two sticks or crutches
Stage IV	Confined to a wheelchair or bedridden

Appendix 2 - FAP Stage Severity Scoring System

FAP Stage	
Stage 0	No symptoms
Stage I	Unimpaired ambulation; mostly mild sensory, motor, and autonomic neuropathy in the lower limbs
Stage II	Assistance with ambulation required; mostly moderate impairment progression to the lower limbs, upper limbs, and trunk
Stage III	Wheelchair bound or bedridden; severe sensory, motor, and autonomic involvement of all limbs

Appendix 3 - List of PA Medications for Polyneuropathy caused by hATTR Amyloidosis

Generic Name	Brand Name
inotersen	Tegsedi
patisiran	Onpattro
eplontersen	Wainua
vutrisiran	Amvuttra