09-J3000-38

Original Effective Date: 06/15/19

Reviewed: 12/11/24

Revised: 01/15/25

Subject: Riluzole (Tiglutik®, Exservan™)

THIS MEDICAL COVERAGE GUIDELINE IS NOT AN AUTHORIZATION, CERTIFICATION, EXPLANATION OF BENEFITS, OR A GUARANTEE OF PAYMENT, NOR DOES IT SUBSTITUTE FOR OR CONSTITUTE MEDICAL ADVICE. ALL MEDICAL DECISIONS ARE SOLELY THE RESPONSIBILITY OF THE PATIENT AND PHYSICIAN. BENEFITS ARE DETERMINED BY THE GROUP CONTRACT, MEMBER BENEFIT BOOKLET, AND/OR INDIVIDUAL SUBSCRIBER CERTIFICATE IN EFFECT AT THE TIME SERVICES WERE RENDERED. THIS MEDICAL COVERAGE GUIDELINE APPLIES TO ALL LINES OF BUSINESS UNLESS OTHERWISE NOTED IN THE PROGRAM EXCEPTIONS SECTION.

Dosage/ Administration	Position Statement	Billing/Coding	Reimbursement	Program Exceptions	Definitions
Related Guidelines	Other	References	Updates		

DESCRIPTION:

Approximately 12,000 to 15,000 individuals in the United States have amyotrophic lateral sclerosis (ALS). Also known as Lou Gehrig's disease, ALS is a progressive, neurodegenerative disease that attacks neurons controlling voluntary muscles. While symptom onset is gradual, those with ALS eventually lose the ability to walk, talk, eat, and ultimately, breathe without ventilator support. The majority of patients with ALS die within 3 to 5 years of developing symptoms. Currently, there is no cure for ALS and treatment is limited to symptom control and supportive therapies. Riluzole and edaravone are the only FDA approved treatments that have shown to be modestly effective in ALS. Riluzole is currently recommended by the American Academy of Neurology for use in ALS to slow the disease process. It is available as an oral tablet, oral film, and oral suspension.

POSITION STATEMENT:

Comparative Effectiveness

The FDA has deemed the drug(s) or biological product(s) in this coverage policy to be appropriate for self-administration or administration by a caregiver (i.e., not a healthcare professional). Therefore, coverage (i.e., administration) in a provider-administered setting such as an outpatient hospital, ambulatory surgical suite, physician office, or emergency facility is not considered medically necessary.

NOTE: Generic riluzole tablets do not require prior authorization.

Initiation of riluzole (Tiglutik, Exservan) **meets the definition of medical necessity** when **ALL** of the following criteria met:

1. Member is diagnosed with amyotrophic lateral sclerosis (ALS)
2. Member is unable to ingest an oral tablet due to oral or motor difficulties or dysphagia – documentation from the medical record must be submitted

3. Member did not achieve treatment goals (such as, treatment failure), had persistent intolerable adverse effects, or has a contraindication to treatment with riluzole tablets
4. Dose does not exceed 100 mg (20 mL or 2 films) daily
5. Dispensed quantity does not exceed 600 mL/30 days or 60 films/30 days

Approval duration: 6 months

Continuation of riluzole (Tiglutik, Exservan) **meets the definition of medical necessity** when **ALL** of the following criteria are met:

1. Authorization/reauthorization has been previously approved by Florida Blue or another health plan in the past two years for ALS, OR the member has previously met all indication-specific initiation criteria
2. Member is unable to ingest an oral tablet due to oral or motor difficulties or dysphagia – documentation from the medical record must be submitted
3. Dose does not exceed 100 mg (20 mL or 2 films) daily
4. Dispensed quantity does not exceed 600 mL/30 days or 60 films/30 days

Approval duration: 6 months

DOSAGE/ADMINISTRATION:

THIS INFORMATION IS PROVIDED FOR INFORMATIONAL PURPOSES ONLY AND SHOULD NOT BE USED AS A SOURCE FOR MAKING PRESCRIBING OR OTHER MEDICAL DETERMINATIONS. PROVIDERS SHOULD REFER TO THE MANUFACTURER'S FULL PRESCRIBING INFORMATION FOR DOSAGE GUIDELINES AND OTHER INFORMATION RELATED TO THIS MEDICATION BEFORE MAKING ANY CLINICAL DECISIONS REGARDING ITS USAGE.

FDA-approved

- 50 mg (10 mL or 1 film) twice daily every 12 hours

Dose Adjustments

- Measure serum aminotransferases before and during treatment
- See FDA-approved product information for additional information

Drug Availability

- Oral suspension: 50 mg/10 mL (5 mg/mL) in 300 mL multiple-dose bottle
- Oral film: 50 mg

PRECAUTIONS:

Boxed Warning

- None

Contraindications

- Patients with a history of severe hypersensitivity reactions to riluzole or to any of its components

Precautions/Warnings

- Hepatic injury: Use is not recommended in patients with baseline elevations of serum aminotransferases greater than 5 times the upper limit of normal; discontinue if there is evidence of liver dysfunction
- Neutropenia: Advise patients to report any febrile illness
- Interstitial lung disease: Discontinue if interstitial lung disease develops

BILLING/CODING INFORMATION:

HCPCS Coding

J8499	Prescription drug, oral, non-chemotherapeutic, Not Otherwise Specified

ICD-10 Diagnosis Codes That Support Medical Necessity

G12.21	Amyotrophic lateral sclerosis

REIMBURSEMENT INFORMATION:

Refer to section entitled **POSITION STATEMENT**.

PROGRAM EXCEPTIONS:

Federal Employee Program (FEP): Follow FEP guidelines.

State Account Organization (SAO): Follow SAO guidelines.

Medicare Part D: BCBSF has delegated to Prime Therapeutics authority to make coverage determinations for the Medicare Part D services referenced in this guideline.

Medicare Advantage: No National Coverage Determination (NCD) and/or Local Coverage Determination (LCD) were found at the time of the last guideline review date.

DEFINITIONS:

None

RELATED GUIDELINES:

None

OTHER:

None

REFERENCES:

1. Aquestive Therapeutics. Exservan (riluzole) oral film. 2024. [cited 11/20/24]. In: DailyMed [Internet]. Bethesda (MD): National Library of Medicine. Available from: https://dailymed.nlm.nih.gov/dailymed/drugInfo.cfm?setid=0a4ad0db-ce67-4724-91d2-88258c2cc9f8
2. Clinical Pharmacology [Internet]. Tampa (FL): Gold Standard, Inc.; 2024 [cited 11/20/24]. Available from: http://www.clinicalpharmacology.com/.
3. ClinicalTrials.gov [Internet]. Bethesda (MD): National Library of Medicine; 2000 Feb 29 - [cited 11/25/21]. Available from: http://clinicaltrials.gov/.
4. DRUGDEX® System [Internet]. Greenwood Village (CO): Thomson Micromedex; Updated periodically [cited 11/20/24].
5. Itf Pharma. Tiglutik (riluzole) liquid. 2024. [cited 11/20/24]. In: DailyMed [Internet]. Bethesda (MD): National Library of Medicine. Available from: https://dailymed.nlm.nih.gov/dailymed/drugInfo.cfm?setid=75d68f19-4208-109e-e053-2991aa0ac857
6. Orphan Drug Designations and Approval [Internet]. Silver Spring (MD): US Food and Drug Administration; 2024 [cited 11/20/24]. Available from: http://www.accessdata.fda.gov/scripts/opdlisting/oopd/index.cfm/.

COMMITTEE APPROVAL:

This Medical Coverage Guideline (MCG) was approved by the Florida Blue Pharmacy Policy Committee on 12/11/24.

GUIDELINE UPDATE INFORMATION:

06/15/19	New Medical Coverage Guideline.
01/15/20	Review and revision of guideline; updated references.
09/15/21	Updated position statement with new formulation.
01/15/22	Review and revision of guideline; updated position statement and references
01/15/25	Review and revision to guidelines consisting of updating the references.