

riluzole (Tiglutik™)

Policy # 00655

Original Effective Date: 12/19/2018

Current Effective Date: 10/01/2026

Applies to all products administered or underwritten by Blue Cross and Blue Shield of Louisiana and its subsidiary, HMO Louisiana, Inc. (collectively referred to as the "Company"), unless otherwise provided in the applicable contract. Medical technology is constantly evolving, and we reserve the right to review and update Medical Policy periodically.

Investigational or experimental services are not covered. This includes any drug, device, procedure, or service provided under the investigational arm of a clinical trial or clinical study. These services are excluded from coverage under benefits.

When Services May Be Eligible for Coverage

Coverage for eligible medical treatments or procedures, drugs, devices or biological products may be provided only if:

- *Benefits are available in the member's contract/certificate, and*
- *Medical necessity criteria and guidelines are met.*

Based on review of available data, the Company may consider riluzole oral suspension (Tiglutik™)‡ for the treatment of amyotrophic lateral sclerosis (ALS) to be **eligible for coverage**. **

Patient Selection Criteria

Coverage eligibility for riluzole oral suspension (Tiglutik) will be considered when ALL of the following criteria are met:

- Patient has a diagnosis of amyotrophic lateral sclerosis (ALS); AND
- Patient has a gastrostomy tube (G-tube) or is otherwise unable to swallow tablets (e.g. has dysphagia or difficulty swallowing tablets); AND
*(Note: This specific patient selection criterion is an additional Company requirement for coverage eligibility and will be denied as not medically necessary** if not met.)*
- Patient is NOT taking any medication in tablet or capsule form.
*(Note: This specific patient selection criterion is an additional Company requirement for coverage eligibility and will be denied as not medically necessary** if not met.)*

When Services Are Considered Not Medically Necessary

Based on review of available data, the Company considers the use of riluzole oral suspension (Tiglutik) when the patient is able to swallow tablets or is currently taking medications in tablet or capsule form to be **not medically necessary**.**

When Services Are Considered Investigational

Coverage is not available for investigational medical treatments or procedures, drugs, devices or biological products.

Based on review of available data, the Company considers the use of riluzole oral suspension (Tiglutik) for indications other than ALS to be **investigational**.*

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Background/Overview

Riluzole is the only known drug to have any impact on survival in amyotrophic lateral sclerosis (ALS). It exerts this activity through an unknown mechanism but is thought to reduce glutamate-induced excitotoxicity. Riluzole is available as brand Rilutek® tablets, generic riluzole tablets, and Tiglutik oral suspension. Because ALS can cause dysphagia and difficulty swallowing, Tiglutik addresses an unmet need to provide a formulation that does not require the patient to swallow tablets or capsules. Tiglutik can be administered orally or via a G tube. It's dosed at 50mg twice daily and should be taken at least 1 hour before or 2 hours after a meal.

ALS is a rapidly progressing, degenerative disease in which the patient's upper and lower motor neurons degenerate leading to loss of motor function. Patients with ALS present with painless, progressive muscle atrophy and weakness, which eventually leads to paralysis. Death due to respiratory failure typically occurs within 3-5 years of diagnosis. Approximately 14,000-15,000 people in the U.S. have ALS. The disease occurs most commonly in people aged 55-75 years. Current treatment guidelines from the American Academy of Neurology as well as guidelines from the European Federation of Neurological Societies recommend riluzole to all patients as early as possible after diagnosis unless the risk of fatigue outweighs the modest survival benefits with the drug.

FDA or Other Governmental Regulatory Approval

U.S. Food and Drug Administration (FDA)

Tiglutik was approved in September 2018 for the treatment of amyotrophic lateral sclerosis (ALS).

Rationale/Source

This medical policy was developed through consideration of peer-reviewed medical literature generally recognized by the relevant medical community, U.S. Food and Drug Administration approval status, nationally accepted standards of medical practice and accepted standards of medical practice in this community, technology evaluation centers, reference to regulations, other plan medical policies, and accredited national guidelines.

Tiglutik was approved based upon bioavailability studies comparing oral riluzole tablets to the new formulation. Clinical efficacy studies were only done with riluzole oral tablets. The patient selection criteria presented in this policy take into consideration whether or not the patient can swallow oral riluzole tablets. Based on a review of the data, if the above-mentioned criteria are not met there is no advantage to the use of Tiglutik over the generic oral riluzole tablets.

References

1. Tiglutik [package insert]. ITF Pharma, Inc. Berwyn, PA. Updated April 2021.
2. Miller RG, Jackson CE, Kasarskis EJ, et al. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: drug, nutritional, and respiratory therapies (an evidence-based review). *Neurology*. 2009;73:1218-1226.

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3. Andersen PM, Abrahams S, Borasio GD, et al. EFNS guidelines on the clinical management of amyotrophic lateral sclerosis (MALS)—revised report of an EFNS task force. *Eur J Neurol.* 2012;19(3):360-375.
4. Neurology-Riluzole Products PA Policy. Express Scripts. Updated December 2019.

Policy History

Original Effective Date: 12/19/2018

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12/06/2018	Medical Policy Committee review
12/19/2018	Medical Policy Implementation Committee approval. New policy.
12/05/2019	Medical Policy Committee review
12/11/2019	Medical Policy Implementation Committee approval. Coverage eligibility unchanged.
12/03/2020	Medical Policy Committee review
12/09/2020	Medical Policy Implementation Committee approval. Coverage eligibility unchanged
09/02/2021	Medical Policy Committee review
09/08/2021	Medical Policy Implementation Committee approval. Added new product, Exservan, to policy with relevant background information.
09/01/2022	Medical Policy Committee review
09/14/2022	Medical Policy Implementation Committee approval. Coverage eligibility unchanged.
09/07/2022	Medical Policy Committee review
09/13/2022	Medical Policy Implementation Committee approval. Coverage eligibility unchanged.
09/05/2024	Medical Policy Committee review
09/11/2024	Medical Policy Implementation Committee approval. Coverage eligibility unchanged.
09/04/2025	Medical Policy Committee review
09/10/2025	Medical Policy Implementation Committee approval. Coverage eligibility unchanged.
09/09/2026	Pre Pharmacy & Therapeutics Committee review
09/16/2026	Pharmacy & Therapeutics Committee review approval. Removed discontinued product, Exservan, from the policy. Title changed from “riluzole (Tiglutik™, Exservan™)” to “riluzole (Tiglutik™)”. Coverage eligibility of remaining product unchanged.

Next Scheduled Review Date: 09/2027

*Investigational – A medical treatment, procedure, drug, device, or biological product is Investigational if the effectiveness has not been clearly tested and it has not been incorporated into standard medical practice. Any determination we make that a medical treatment, procedure, drug, device, or biological product is Investigational will be based on a consideration of the following:

- A. Whether the medical treatment, procedure, drug, device, or biological product can be lawfully marketed without approval of the U.S. Food and Drug Administration (FDA) and whether such approval has been granted at the time the medical treatment, procedure, drug, device, or biological product is sought to be furnished; or

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- B. Whether the medical treatment, procedure, drug, device, or biological product requires further studies or clinical trials to determine its maximum tolerated dose, toxicity, safety, effectiveness, or effectiveness as compared with the standard means of treatment or diagnosis, must improve health outcomes, according to the consensus of opinion among experts as shown by reliable evidence, including:
1. Consultation with technology evaluation center(s);
 2. Credible scientific evidence published in peer-reviewed medical literature generally recognized by the relevant medical community; or
 3. Reference to federal regulations.

****Medically Necessary (or “Medical Necessity”)** - Health care services, treatment, procedures, equipment, drugs, devices, items or supplies that a Provider, exercising prudent clinical judgment, would provide to a patient for the purpose of preventing, evaluating, diagnosing or treating an illness, injury, disease or its symptoms, and that are:

- A. In accordance with nationally accepted standards of medical practice;
- B. Clinically appropriate, in terms of type, frequency, extent, level of care, site and duration, and considered effective for the patient's illness, injury or disease; and
- C. Not primarily for the personal comfort or convenience of the patient, physician or other health care provider, and not more costly than an alternative service or sequence of services at least as likely to produce equivalent therapeutic or diagnostic results as to the diagnosis or treatment of that patient's illness, injury or disease.

For these purposes, “nationally accepted standards of medical practice” means standards that are based on credible scientific evidence published in peer-reviewed medical literature generally recognized by the relevant medical community, Physician Specialty Society recommendations and the views of Physicians practicing in relevant clinical areas and any other relevant factors.

‡ Indicated trademarks are the registered trademarks of their respective owners.

NOTICE: If the Patient’s health insurance contract contains language that differs from the BCBSLA Medical Policy definition noted above, the definition in the health insurance contract will be relied upon for specific coverage determinations.

NOTICE: Medical Policies are scientific based opinions, provided solely for coverage and informational purposes. Medical Policies should not be construed to suggest that the Company recommends, advocates, requires, encourages, or discourages any particular treatment, procedure, or service, or any particular course of treatment, procedure, or service.

NOTICE: Federal and State law, as well as contract language, including definitions and specific contract provisions/exclusions, take precedence over Medical Policy and must be considered first in determining eligibility for coverage.

NOTICE: If an authorization for an ongoing course of treatment has been provided to a member and the member changes from one health plan to another health plan (e.g., a member moves from carrier A to Louisiana Blue), Louisiana Blue may honor the previous health plan’s authorization for the same service under the same type of in-network benefit for a 90-day transition period.

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Documentation of the authorization for the ongoing course of treatment from the previous health plan must be provided to us by the member or their provider and the services provided for the course of treatment must otherwise be a covered service under the Louisiana Blue health plan. This provision does not apply to medications covered under the plan's pharmacy benefit.