PRIOR AUTHORIZATION POLICY

POLICY: Neurology – Riluzole Products Prior Authorization Policy
- Exservan™ (riluzole oral film – Covis Pharmaceuticals/Aquestive)
- Rilutek® (riluzole tablets – Covis Pharma; generics)
- Tiglutik® (riluzole oral suspension – ITF Pharma)

REVIEW DATE: 07/22/2020

OVERVIEW

All of the available riluzole products are indicated for the treatment of amyotrophic lateral sclerosis (ALS).[1,2,9] Riluzole is a member of the benzothiazole class; the mechanism by which it exerts its effects in patients with ALS is unknown. Riluzole tablets were initially approved by the FDA in 1995.[1] In the years since, two additional riluzole formulations, Exservan oral film and Tiglutik oral suspension, have been approved.[2,9]

Disease Overview

ALS, also known as Lou Gehrig's disease, is a rapidly progressive, paralyzing disease characterized by degeneration of upper motor neurons (UMNs) and lower motor neurons (LMNs) in the brain, brainstem, and the spinal cord resulting in muscle weakness.[3-5] Patients with ALS typically present with painless, progressive muscle atrophy and weakness, which eventually leads to paralysis and death, primarily due to respiratory failure. The accurate diagnosis of ALS is challenging and delays in diagnosis are common (average diagnostic delay of 11 to 12 months or more). The average survival following diagnosis of ALS is approximately 3 years; 50% of patients will die within 30 months of symptom onset. However, median survival can range from months to several years and the rate of progression of the disease varies considerably between patients.

Guidelines

The American Academy of Neurology (AAN) practice parameter on the care of patients with ALS (last updated 2009; reaffirmed 2020) states that riluzole should be offered to patients with ALS (Level A recommendation), as it is safe and effective for modestly slowing disease progression.[6,7] Based on available clinical trial data, the AAN estimates riluzole prolongs survival by 2 to 3 months. However, some large cohort studies that estimate survival to be prolonged for up to 21 months. Of note, a previous practice advisory from the AAN (1997) had recommended riluzole only in patients with definite or probable ALS of < 5 years duration, with a forced vital capacity (FVC) > 60% and without tracheostomy. However, the 2012 parameter does not include any stipulations on which patients should use riluzole. Riluzole may result in fatigue in some patients and if the risk of fatigue outweighs modest survival benefits, discontinuation of riluzole may be considered (Level C recommendation). The European Federation of Neurological Societies (EFNS) guidelines on the clinical management of ALS (2012) also recommend patients be offered treatment with riluzole as early as possible after diagnosis.[8] While it is noted that riluzole may be less effective in patients with late-stage disease, it is unclear when or if treatment should be discontinued. Patients with progressive muscular atrophy, primary lateral sclerosis, or hereditary spastic paraplegia are not recommended to use riluzole as they were not included in the clinical studies. However, some of these patients fall within the ALS syndrome and therefore may benefit from treatment with riluzole.

POLICY STATEMENT

Prior authorization is recommended for prescription benefit coverage of riluzole. All approvals are provided for the duration noted below. Because of the specialized skills required for evaluation and diagnosis of patients treated with riluzole as well as the monitoring required for adverse events and long-term efficacy, approval requires riluzole to be prescribed by or in consultation with a physician who specializes in the condition being treated.

Automation: None.

RECOMMENDED AUTHORIZATION CRITERIA

Coverage of riluzole is recommended in those who meet the following criteria:

FDA-Approved Indications

1. **Amyotrophic Lateral Sclerosis (ALS).** Approve for 1 year if the agent is prescribed by or in consultation with a neurologist, a neuromuscular disease specialist, or a physician specializing in the treatment of ALS.

CONDITIONS NOT RECOMMENDED FOR APPROVAL

Coverage of riluzole is not recommended in the following situations:

1. Coverage is not recommended for circumstances not listed in the Recommended Authorization Criteria. Criteria will be updated as new published data are available.

REFERENCES

1. Rilutek tablets [prescribing information]. Zug, Switzerland: Covis PharmaB.V.; March 2020.
2. Tiglutik® oral suspension [prescribing information]. Berwyn, PA: ITF Pharma, Inc. March 2020.
3. Petrolv D, Mansfield C, Moussy A, et al. ALS clinical trials review: 20 years of failure. Are we any closer to registering a new treatment? *Front Aging Neurosci*. 2017;9:68.
4. Salameh JS, Brown RH, Berry JD. Amyotrophic lateral sclerosis: review. *Semin Neurol*. 2015;35(4):469-476.
5. Kiernan MC, Vucic S, Cheah BC, et al. Amyotrophic lateral sclerosis. *Lancet*. 2011;377:942-955.
6. Miller RG, Jackson CE, Kasarskis EJ, et al. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: multidisciplinary care, symptom management, and cognitive/behavioral impairment (an evidence-based review). *Neurology*. 2009;73(15):1227-1233.
7. 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.
8. 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.
9. Exservan™ sublingual film [prescribing information]. Warren, NJ: Aquestive Therapeutics. May 2020.