ITF PHARMA ANNOUNCES FDA APPROVAL OF SUPPLEMENTAL NEW DRUG APPLICATION (sNDA) FOR TIGLUTIK® (RILUZOLE) ORAL SUSPENSION FOR THE TREATMENT OF AMYOTROPHIC LATERAL SCLEROSIS (ALS) WITH A PERCUTANEOUS ENDOSCOPIC GASTROSTOMY (PEG) FEEDING TUBE

~ TIGLUTIK is the only formulation of riluzole indicated for both oral and PEG tube administration ~

Berwyn, Pa., Dec. 13, 2019 – ITF Pharma, a U.S.-based specialty pharmaceutical company and subsidiary of Italfarmaco, a privately-held European specialty pharmaceutical company, announced today that the U.S. Food and Drug Administration (FDA) has approved its application to broaden the existing label for TIGLUTIK® (riluzole) oral suspension to include administration via percutaneous endoscopic gastrostomy (PEG) tubes for the treatment of amyotrophic lateral sclerosis (ALS). This supplemental new drug application (sNDA) approval expands the patient population who stand to benefit from TIGLUTIK to include individuals whose swallowing difficulty is complicated by alterations in nutritional status, necessitating the use of a PEG feeding tube.

Approximately 20,000 Americans are living with ALS.[i] Difficulty swallowing, medically referred to as dysphagia, afflicts 85 percent of people with ALS during the course of their disease.[ii] Due to these challenges, many patients require the placement of a PEG tube, which allows nutrition, fluids and medications to be put directly into the stomach, bypassing the mouth and esophagus.

“There are many medical and quality of life advantages for the use of a PEG feeding tube as an individual’s ALS progresses, including the ability maintain more adequate hydration, administer medications and to potentially reduce the risk of choking and aspirating when oral ingestion becomes too challenging,” said Benjamin Rix Brooks, M.D., medical director at the Carolinas Neuromuscular/ALS - MDA Center. “This expanded use for TIGLUTIK provides an alternate administration route for the majority of people with ALS who choose to undergo this procedure.”

The American Academy of Neurology (AAN) recommends an evaluation of the nutritional status of ALS patients every three months and advises consideration of feeding tube placement when patients demonstrate swallowing difficulty or alterations in nutritional status.[iii]

“We are very pleased the FDA has approved the application to broaden the existing label for TIGLUTIK to include PEG tube administration. This news marks another important step forward in the treatment of ALS and addresses an unmet treatment need of the ALS community,” said Peter Cook, president and chief executive officer of ITF Pharma.

TIGLUTIK, initially approved in 2018, is an oral suspension formulation of riluzole which helps fulfill an important therapeutic unmet need for the approximately 85 percent of ALS patients who develop dysphagia. Riluzole has been the gold standard of treatment since 1995 to slow the progression of ALS, an ultimately fatal neurodegenerative disease that still has no cure. TIGLUTIK has received orphan drug designation from the FDA.

This FDA approval of TIGLUTIK is based on a study that showed TIGLUTIK was bioequivalent when administered intragastrically and orally.[iv] The most common side effects of TIGLUTIK are consistent with the established clinical profile of riluzole and include oral hypoesthesia, asthenia, nausea, decreased lung function, hypertension and abdominal pain. While the mechanism of action of riluzole is not fully understood, in clinical studies it has been shown repeatedly to modulate glutamate neurotransmission by inhibiting both glutamate release and postsynaptic glutamate receptor signaling.

About Amyotrophic Lateral Sclerosis (ALS)
Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease, is a progressive, ultimately fatal neurodegenerative disease, marked by a gradual degeneration of nerve cells of the central nervous system that control voluntary muscle movement.[v] According to the ALS Association and based on U.S. population studies, a little over 5,000 people in the U.S. are diagnosed with ALS each year. It is estimated that 20,000 Americans have the disease at any given time. The incidence of ALS increases with age, typically starting in the 40s and continuing until around the age of 80. However, ALS can occur in people in their 20s and 30s.[vi] In ALS, the degeneration of motor neurons is characterized by muscle weakness, typically impacting arms and legs, speech, swallowing and breathing. Impairment of swallowing (dysphagia) is a feature of ALS resulting from weakness or spasticity of muscles affecting the tongue, lips, palate, jaw, pharynx, larynx and upper trunk.[vi]

About TIGLUTIK® (riluzole) Oral Suspension
TIGLUTIK is the only formulation of riluzole indicated for both oral and PEG tube administration. It is administered twice daily by either an oral syringe or using a PEG tube. In clinical studies, the most common side effects of TIGLUTIK were oral hypoesthesia, asthenia, nausea, decreased lung function, hypertension and abdominal pain. These are not all of the possible side effects that you may experience with TIGLUTIK. Talk to your doctor if you have any symptoms that bother you or do not go away.

Indication
TIGLUTIK is indicated for the treatment of amyotrophic lateral sclerosis (ALS).

Important Safety Information
- You should not take TIGLUTIK if you are allergic to any of its ingredients.
- TIGLUTIK can cause liver injury, including death. Your doctor should do blood tests to check your liver function before and during your treatment and may stop treatment with TIGLUTIK if liver function is not normal. Contact your doctor immediately if you have unexplained nausea, vomiting, abdominal pain, fatigue, anorexia, or jaundice and/or dark urine
- Call your doctor immediately if you have a fever, cough, or difficulty in breathing while taking TIGLUTIK.
- If you miss or skip a dose of TIGLUTIK, do not take any extra doses to make up for those you missed, but take your prescribed dose at the next regularly scheduled time.
- The most common side effects of TIGLUTIK that occurred during medical studies were numbness/tingling around the mouth, weakness, nausea, decreased lung function, high blood pressure and abdominal pain. If any side effects become troublesome, contact your doctor.
- Be sure to tell your doctor and pharmacist about all other health conditions you have and all medicines you are taking, including nonprescription products and vitamins. If you have questions, please talk to your doctor.

You are encouraged to report negative side effects of prescription drugs to the FDA. Visit www.fda.gov/medwatch, or call 1-800-332-1088.

Please click here for the full Prescribing Information.

About ITF Pharma
ITF Pharma (www.itfpharma.com) is a Pennsylvania-based, specialty pharmaceutical company committed to investing in and commercializing impactful medicines in therapeutic areas with unfulfilled needs. ITF Pharma is the U.S. subsidiary of Italfarmaco, a privately held European specialty pharmaceutical company engaged in the development of new and groundbreaking therapies. ITF's commercial portfolio includes TIGLUTIK® (riluzole) oral suspension, the first and only thickened liquid formulation of riluzole, approved for the treatment of amyotrophic lateral sclerosis (ALS) by the U.S. Food and Drug Administration in September of 2018.

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[ii] Onesti, E., Schettino, I., Gori, M.C. et al. Dysphagia in amyotrophic lateral sclerosis: impact on patient behavior, diet adaptation, and riluzole management. (eCollection 2017) *Front Neurol*. 2017 Mar 21; 8: 94https://doi.org/10.3389/fneur.2017.00094.
[iii] Miller RG, Rosenberg JA, Gelinas DF, et al; ALS Practice Parameters Task Force. Practice parameter: The care of the patient with amyotrophic lateral sclerosis (an evidence-based review): Report of the Quality Standards Committee of the American Academy of Neurology. *Neurology*. 1999;52(7):1311-1323.
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[v] AM Dyer and A Smith. Riluzole 5 mg/mL oral suspension: for optimized drug delivery in amyotrophic lateral sclerosis. *Drug Des Devel Ther*. 2017; 11: 59–64.
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