NEWS RELEASE

Prilenia Receives Fast Track Designation for Pridopidine for the Treatment of Huntington's Disease

11/17/2021

-- Fast Track designation may accelerate the registration process for pridopidine by providing the ability to file a rolling NDA and qualify for priority review --

NAARDEN, Netherlands--(**BUSINESS WIRE**)--**Prilenia Therapeutics B.V.**, a clinical stage biotech company focused on developing novel treatments for neurodegenerative and neurodevelopmental disorders, today announced that the U.S. Food and Drug Administration (FDA) has granted Fast Track designation to pridopidine for development as a potential treatment for **Huntington's Disease** (HD).

Fast Track is a process designed to facilitate the development and expedite the review of new treatments for serious conditions with unmet medical need such as HD. Drugs that receive Fast Track designation may be eligible for more frequent communications with the FDA to review the drug's development plan, including the extent of data needed for approval. Drugs with Fast Track designation may also qualify for accelerated approval and priority review of new drug applications.

Pridopidine is currently being assessed in **PROOF-HD** (PRidopidine Outcome On Function in Huntington Disease), a global phase 3 clinical trial that very recently completed patient enrollment. Pridopidine acts as a highly selective and potent Sigma-1 Receptor (S1R) agonist.

“Receipt of Fast Track designation from the FDA underscores the urgency to address a significant unmet need for patients with Huntington’s Disease,” said Dr. Michael R. Hayden, CEO and Founder of Prilenia. “HD is one of the most devastating neurodegenerative disorders, impacting not only patients but their families. At the present time, there is no approved treatment to delay onset or slow disease progression. We look forward to working closely with the FDA in making pridopidine a potential option for these patients.”

“The FDA Fast Track designation is an important milestone, as it provides opportunities to work collaboratively with the FDA to accelerate pridopidine’s development for the treatment of Huntington’s Disease,” said Henk Schuring, Chief Regulatory and Commercialization Officer at Prilenia.

This designation follows Prilenia’s recent company announcements, including the **PROOF-HD completion of enrollment ahead of schedule** and the **closing of series B financing round**.

About Pridopidine

Prilenia’s lead asset, **pridopidine** is a highly selective and potent Sigma-1 Receptor (S1R) agonist with an established safety and tolerability profile. Pridopidine is orally administered with therapeutic potential in HD, ALS and other neurodegenerative diseases and neurodevelopmental disorders such as Rett and Fragile-X syndromes.

Pridopidine is currently in late-stage clinical development for HD and ALS. Both trials, the global phase 3 clinical trial in HD (PROOF-HD) and the Healey platform trial in ALS are currently active.

About Prilenia

Prilenia is a clinical stage biotech startup founded in 2018 with the purpose of improving the lives of patients and their families by developing treatments for neurodegenerative and neurodevelopmental disorders. Prilenia is backed by a group of well-respected investors including: Forbion, Morningside, Sands Capital, Sectoral Asset Management, Talisman, Amplitude Ventures and the ALS Investment Fund. The Company is based in Naarden, the Netherlands, Herzliya, Israel and Boston, MA in the U.S. For more information visit **www.prilenia.com** and follow us on Twitter **@prileniaTx**.

Pridopidine for Huntington’s Disease

Huntington’s Disease (HD) is a fatal, inherited, neurodegenerative disorder. Every offspring of an HD patient has a 50% chance of inheriting the gene. Usually starting at around 40 years of age, HD patients suffer from a movement disorder, progressive functional and cognitive decline, psychiatric disturbances and behavioral symptoms. Following diagnosis, functional, motor and cognitive functions declines, ultimately leading to immobility, dementia

and premature death.

Pridopidine has demonstrated maintenance of functional capacity in HD patients, as measured by Total Functional Capacity (TFC), in a post hoc analysis of a phase 2 clinical trial. This effect was most prominent in early-stage HD patients (HD1 and HD2), who showed functional benefit from pridopidine 45 mg, taken twice a day.

Pridopidine demonstrates neuroprotective properties mediated by the S1R in several in vivo and in vitro HD models. In all these models, when the S1R is deleted or an S1R antagonist is used, these neuroprotective properties are eliminated completely. These are comprised of a robust, neuroprotective effect against mutant huntingtin-(mHTT)-induced cell death in human HD induced pluripotent stem cells (iPSCs), and mouse HD cortical neurons. In HD cortico-striatal cultures, pridopidine increases spine density and rescues the aberrant calcium signaling, enhances mitochondrial function and mitigates mHtt-induced ER and oxidative stress, all known features of HD.

Prilenia has been granted orphan drug designation for pridopidine for the treatment of HD in both the U.S. and Europe.

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