NEWS RELEASE

Prilenia Plans to Submit Marketing Authorization Application (MAA) in the EU for Pridopidine in Huntington's Disease

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-Following positive pre-submission meetings with regulators, Prilenia plans to submit its MAA for pridopidine in Huntington's disease (HD) in mid-2024-

-If approved, pridopidine could be commercially available to patients in Europe as early as 2025-

NAARDEN, Netherlands & WALTHAM, Mass.--(BUSINESS WIRE)-- **Prilenia Therapeutics B.V.**, a clinical stage biotechnology company focused on the urgent mission to develop novel therapeutics to slow the progression of neurodegenerative diseases and neurodevelopmental disorders, announced its plan to submit a Marketing Authorization Application (MAA) for pridopidine for the treatment of Huntington's disease (HD) to the European Medicines Agency's (EMA) Committee for Medicinal Products for Human Use (CHMP). This decision follows positive pre-submission meetings with regulatory authorities in the European Union. Submission is planned for mid-2024.

"Pridopidine demonstrates consistent treatment benefits across independent measures that are important to patients and families. These measures include day-to-day function, cognition, motor and clinical progression in people living with HD. The benefits are clearly evident in those with HD who are not taking anti-dopaminergic medications (ADM)," said Dr. Michael R. Hayden, CEO of Prilenia. "We appreciate the constructive discussions with European regulators regarding our data for pridopidine. This provides a pathway to potential approval of a therapy for HD, a rare neurodegenerative disease with a predictable decline, and no currently approved treatments that

address progression. We are now finalizing our MAA submission for mid-2024 and preparing to make pridopidine commercially available for patients in Europe, if approved."

ADMs include neuroleptics (also known as antipsychotics) and VMAT2 inhibitor (anti-chorea) drugs.

Prilenia intends to discuss with the U.S. Food and Drug Administration (FDA) a potential path forward for pridopidine as a possible treatment for those living with HD in the United States. The Company will also consider global regulatory submissions for additional countries and regions following the regulatory review process in Europe.

In addition to the progress in HD, Prilenia **announced** in January 2024 that it had completed discussions with global regulatory agencies regarding the next stage of development of pridopidine for amyotrophic lateral sclerosis (ALS) and is planning for a single pivotal Phase 3 study.

About Pridopidine

Pridopidine (45 mg twice daily) is an oral, highly selective and potent investigational S1R agonist that has exhibited a safety and tolerability profile similar to placebo in clinical studies to date. The safety dataset includes approximately 1,700 patients who have been on pridopidine for up to seven years. The S1R protein is highly expressed in the brain and spinal cord, where it regulates several key processes that are commonly impaired in various neurodegenerative diseases. Activation of the S1R by pridopidine stimulates multiple cellular protective pathways including enhancing autophagy, axonal transport, mitochondrial energy production and respiration, and restores calcium homeostasis. These effects are essential to neuronal function and survival, and lead to neuroprotective effects.

Prilenia holds Orphan Drug designation for pridopidine in HD and ALS in the U.S. and EU. In addition, pridopidine has received Fast Track designation by the U.S. Food and Drug Administration (FDA) for the treatment of HD.

About Huntington's disease

Huntington's disease (HD) is a rare inherited (genetic) neurodegenerative disorder caused by a mutation in the huntingtin gene. Each child of a parent with HD has a 50 percent chance of developing the disease. HD causes neurons in the brain to degenerate and lose their ability to communicate with each other, resulting in functional, motor, cognitive and behavioral symptoms. HD affects about 100,000 people around the world with an additional 300,000 people at risk of developing HD. It is usually diagnosed between the ages of 30 and 50, although HD can also occur at any age including children and young adults (known as juvenile onset HD or JHD). The disease progresses slowly over 15 to 20 years, with patients slowly losing their ability to work, communicate, manage day to

day life and take care of themselves. Currently, there is no treatment to address the progression of HD.

About PROOF-HD

PROOF-HD, a Phase 3, randomized, double-blind, placebo-controlled study, evaluated the safety and efficacy of pridopidine (45 mg twice daily for 65-78 weeks), in individuals with HD (**NCT04556656**). The study enrolled 499 individuals and was conducted in the U.S., Canada, Austria, Czech Republic, France, Germany, Italy, the Netherlands, Poland, Spain and the United Kingdom.

Pridopidine (45 mg bid) has shown consistent and sustained benefit on multiple, independent, clinically meaningful measures in HD participants not taking ADMs. These benefits were seen across multiple clinical endpoints that matter to people with HD and their families (function, cognition, motor, global disease progression and quality of life). In pre-specified analyses, there were improvements compared to placebo across progression, motor, cognition and HD QoL (as measured by TFC, cUHDRS, SWR, Q-Motor, HDQoL). These treatment benefits were sustained for up to 78 weeks at all time points. There was also unprecedented improvement from baseline for a minimum of 1 year in cUHDRS, cognition and motor function (Q-Motor). These PROOF-HD results are further supported by an independent integrated efficacy analysis from a 4 placebo-controlled, double-blind studies of pridopidine in HD which showed significance for cUHDRS, TFC and Q-Motor in patients off ADMs.

In clinical studies to date, pridopidine's safety and tolerability profile was comparable to placebo. The PROOF-HD study was conducted in collaboration with the **Huntington Study Group (HSG)**, a world leader in clinical research for HD and valued collaborator for Prilenia.

About Prilenia

Prilenia is a clinical stage biotechnology company founded in 2018 focused on the urgent mission to develop novel therapeutics to slow the progression of neurodegenerative diseases and neurodevelopmental disorders. The initial focus of the company has been on HD and ALS.

Prilenia is backed by a group of well-respected investors including: Forbion, Morningside, Sands Capital, SV Health Investors, Sectoral Asset Management, Talisman, Amplitude Ventures and the ALS Investment Fund. The Company is based in the Netherlands, Israel and Massachusetts in the U.S.

For more information, visit **www.prilenia.com** and follow us on **LinkedIn** and **Twitter**.

For a copy of this release, visit Prilenia's website at **www.prilenia.com**.

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