AM-PHARMA ANNOUNCES UPDATED CLINICAL DEVELOPMENT STRATEGY

- **Advancing ilofotase alfa into a Phase 2 study for lead indication cardiac surgery-associated renal damage;**
- **Phase 1b study of ilofotase alfa as enzyme replacement therapy in hypophosphatasia patients being finalized, results to be announced before end of 2023;**
- **Juliane Bernholz, AM-Pharma's Chief Operating Officer since 2019, succeeds Erik van den Berg as Chief Executive Officer. Erik van den Berg continues as board member and strategic advisor**

Utrecht, The Netherlands, September 26, 2023 – AM-Pharma B.V. today announced an updated clinical development strategy to evaluate ilofotase alfa in two indications. The company's proprietary compound ilofotase alfa is a recombinant alkaline phosphatase with an extensive clinical record and proven safety profile.

Based on the significant improvements in Major Adverse Kidney Events by day 90 (MAKE90) observed in the company's Phase 2 and Phase 3 studies with ilofotase alfa in sepsis-associated acute kidney injury (AKI), AM-Pharma advances ilofotase alfa as a potential preventive treatment for Cardiac Surgery-Associated Renal Damage (CSA-RD). In addition, based on preclinical data showing improved overall survival and restored phenotypes associated with hypophosphatasia (HPP), the company develops ilofotase alfa as a potential enzyme replacement therapy in HPP.

AM-Pharma also announced the appointment of Juliane Bernholz, Ph.D., as Chief Executive Officer. Juliane previously served as AM-Pharma's Chief Operating Officer. She will succeed Erik van den Berg, Chief Executive Officer since 2011, who will join the company's Supervisory Board and remain as strategic advisor to the company. To align the organization with ongoing activities, AM-Pharma has completed a reduction of its workforce.

"*With the updated development strategy and funding in place for the CSA-RD Phase 2 study and awaiting imminent results from the Phase 1b in HPP, I am excited to pass on the baton to Juliane after serving 12 years as AM-Pharma's CEO,"* **said Erik van den Berg, Supervisory Board member and former Chief Executive Officer of AM-Pharma.** *"Juliane's expertise in clinical and corporate development, together with her experience as AM-Pharma's COO over the past four years, will be extremely useful as the company pursues its development strategy. I am grateful to all at AM-Pharma and the clinical site staff who have contributed to the development of ilofotase alfa from initial studies to the current clinical stage of the program."*

"*Ilofotase alfa has reduced MAKE90 consistently in two large clinical studies involving over 1,000 subjects with sepsis and AKI. These findings together with an outstanding tolerability profile support a potential reno-protective effect, which we aim to demonstrate in the upcoming Phase 2 study evaluating ilofotase alfa in CSA-RD. Together with our recently initiated clinical program studying ilofotase alfa as enzyme replacement therapy in adult patients with HPP, AM-Pharma is focusing on helping patients with severe illness, who are medically underserved, and I am working closely with the AM-Pharma team to achieve this goal,"* **said Juliane Bernholz, Chief Executive Officer of AM-Pharma.**

The planned double-blind, randomized, placebo-controlled Phase 2 clinical trial will evaluate the safety and efficacy of ilofotase alfa in 150 patients, who are at risk of developing renal damage based on pre-operative kidney function, scheduled for complex open-heart surgery performed with the use of a cardiopulmonary bypass pump.

In parallel, the company is currently exploring the potential of ilofotase alfa as an enzyme replacement therapy in HPP, a rare disorder impairing bone metabolism and muscle strength, in a Phase 1b study in adult patients. The company anticipates announcing the first clinical results before the end of 2023.

AM-Pharma's revised clinical strategy is backed by existing investors who have provided additional funding.

"AM-Pharma remains committed to demonstrate the potential of ilofotase alfa for critically ill kidney and HPP patients," **continued Mark Altmeyer, Chairman of the Board at AM-Pharma**. *"We look forward to collaborating with Juliane and thank Erik for his unyielding dedication over the past 12 years and we are excited to have him join the Board."*

About ilofotase alfa

Ilofotase alfa is a proprietary recombinant alkaline phosphatase, constructed from two human isoforms of alkaline phosphatase, that has been shown to be stable and highly active in multiple clinical trials. The recombinant enzyme displays exquisite activity towards dephosphorylating and detoxifying damage-associated molecular patterns (DAMPs) and pathogen-associated molecular patterns (PAMPs) such as lipopolysaccharide (LPS), ATP, ADP and other extracellular substrates that drive acute inflammation, coagulation and microvascular ischemia found in kidney following sepsis or ischemia-induced damage. Research has shown that ATP dephosphorylation has a double effect in protecting against kidney injury. When the pro-inflammatory ATP is dephosphorylated, the resulting adenosine further reduces inflammation through the activation of the immunosuppressive adenosine A2a receptor pathway. In hypophosphatasia, ilofotase alfa addresses elevated levels of pyridoxal-5'-phosphate (PLP), inorganic pyrophosphates (PPi), two disease related biomarkers that are related to, for example, bone mineralization and pain sensation.

About cardiac surgery-associated renal damage and hypophosphatasia

CSA-RD is a clinical complication that arises from acute kidney injury following open heart surgery performed with the use of a cardiopulmonary bypass pump. AKI occurs in about 30% of patients undergoing this surgical procedure, and there are currently no therapeutic options to prevent the long-term renal impairment caused by CSA-RD.

HPP is a rare inherited disease characterized by a deficiency in alkaline phosphatase which is essential for bone mineralization and general functions. HPP patients of all ages can exhibit a wide variety of symptoms that worsen overtime, including bone injury, severe muscle pain and weakness. The disease can also lead to life-threatening complications in infants. The US Food and Drug Administration (FDA) and the European Medicines Agency (EMA) granted AM-Pharma Orphan Drug Designation (ODD) for ilofotase alfa in HPP. In preclinical models, ilofotase alfa was able to achieve improved overall survival of severe disease, as well as restoration of phenotypes associated with HPP.

About AM-Pharma

AM-Pharma strives to develop medicines for patients confronted with severe medical conditions. Our proprietary asset, ilofotase alfa, is being developed for the treatment of patients with acute kidney injury and has been granted FDA fast-track status. We also develop ilofotase alfa in the severe rare disease hypophosphatasia where ilofotase alfa has orphan drug status in the US and EU. With approximately 1,000 subjects evaluated to date in clinical trials, ilofotase alfa has a proven safety profile and a demonstrated kidney benefit. We are a dedicated team driven to bring treatment options to severely ill patients, their families and acute care professionals. Find out more about us online at: www.am-pharma.com.

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