Press Release – No. 21 / 2019

Zealand Pharma initiates second Phase 3 trial with dasiglucagon for the treatment of congenital hyperinsulinism

- **Congenital hyperinsulinism (CHI) is a rare pediatric disease with high unmet medical needs and serious, debilitating lifelong complications**
- **This Phase 3 trial will evaluate dasiglucagon as treatment for CHI children ages 7 days to 1 year who are dependent on intravenous glucose infusions**
- **Dasiglucagon has been granted a Rare Pediatric Disease designation by the FDA for treatment of CHI in November 2019**

Copenhagen, December 4, 2019 – Zealand Pharma A/S ("Zealand") (NASDAQ: ZEAL), a Copenhagen-based biotechnology company focused on the discovery and development of innovative peptide-based medicines, today announced that the second Phase 3 trial to evaluate dasiglucagon as a potential treatment option for children with congenital hyperinsulinism (CHI) has been initiated.

This Phase 3 trial will enroll up to 12 CHI children from the ages of 7 days to 1 year who are dependent upon intravenous glucose infusions to maintain their plasma glucose levels. The aim of the trial is to evaluate the safety and efficacy of dasiglucagon treatment to reduce the intravenous glucose infusion rate, number of hypoglycemic events, and total amount of carbohydrate administered. Details are available at https://clinicaltrials.gov/ct2/show/NCT04172441. An ongoing Phase 3 trial is evaluating the safety and efficacy of dasiglucagon versus current standard treatments to reduce the number of hypoglycemic events in up to 32 children with CHI age 3 months to 12 years. This trial has included 16 children so far, of which 9 have finished and entered the long-term Phase 3 extension trial. Results from this study are expected in 2020. Details are available at https://clinicaltrials.gov/ct2/show/NCT03777176.

For dasiglucagon as a treatment for CHI, the U.S. Food and Drug Administration (FDA) and European Medicines Agency (EMA) granted Orphan Drug designation in 2017, and the FDA granted a Rare Pediatric Disease designation in November 2019. The FDA defines a "rare pediatric disease" as a serious or life-threatening rare disease primarily affecting individuals aged from birth to 18 years.[1] Zealand's program may also be eligible for priority review by the FDA following the submission of a new drug application (NDA).

"Congenital hyperinsulinism puts an extreme burden on the affected children and their families," said **Adam Steensberg, Chief Medical and Development Officer** at Zealand Pharma. "Babies born with CHI face immediate need for intensive hospital treatment, suffer from frequent and dangerous low blood glucose, and require constant feeding. Without effective treatment, some will undergo pancreatic surgery. We aim to reduce these hardships by offering dasiglucagon as a potential future treatment."

[1] FDA.gov, *Rare Pediatric Disease (RPD) Designation Program*

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About dasiglucagon (glucagon analog stable in liquid formulation)
Dasiglucagon is a Zealand-invented glucagon analog with a unique stability profile in a ready-to-use aqueous solution. The molecule has been designed to be stable in solutions and thus suitable for chronic pump use. Both the U.S. FDA and the European Commission granted orphan drug designation in 2017, and the U.S. FDA granted a rare pediatric disease designation in 2019, for dasiglucagon for the treatment of CHI.

Zealand is also pursuing several indications where dasiglucagon's stable profile could provide new treatment options: dasiglucagon HypoPal® rescue pen for severe hypoglycemia and dasiglucagon dual-hormone pump therapy for diabetes management.

About congenital hyperinsulinism
CHI is a rare pediatric disease which affects mainly newborns, infants and toddlers. Due to a genetic defect in the insulin producing cells, these children have too high insulin levels, resulting in persistent low blood sugar throughout childhood. Current treatment options are often insufficient and necessitate surgical intervention through pancreatectomy, a demanding surgery invariably leading to type 1 diabetes development.

About Zealand Pharma A/S
Zealand Pharma A/S (Nasdaq Copenhagen and New York: ZEAL) ("Zealand") is a biotechnology company focused on the discovery and development of innovative peptide-based medicines. More than 10 drug candidates invented by Zealand have advanced into clinical development, of which two have reached the market. Zealand's current pipeline of internal product candidates focus on specialty gastrointestinal and metabolic diseases. Zealand's portfolio also includes two clinical license collaborations with Boehringer Ingelheim and pre-clinical license collaboration with Alexion Pharmaceuticals.

Zealand is based in Copenhagen (Søborg), Denmark. For further information about the Company's business and activities, please visit www.zealandpharma.com or follow Zealand on LinkedIn or Twitter @ZealandPharma.