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Zealand Pharma doses first patient in **Phase 3 trial with dasiglucagon for the treatment of congenital hyperinsulinism**

- **Congenital hyperinsulinism (CHI) is a rare pediatric disease with high unmet medical need**
- **Dasiglucagon has the potential to transform the lives of children with CHI and possibly save them from undergoing major surgery with severe lifelong implications**

Copenhagen, March 6, 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 the dosing of the first patient in the first Phase 3 trial to evaluate dasiglucagon as a potential treatment option for children with CHI.

"With the first child dosed, this Phase 3 trial takes an important step toward establishing dasiglucagon as a potential treatment of congenital hyperinsulinism," said **Adam Steensberg, Interim CEO and Chief Medical and Development Officer** at Zealand. "There is a critical need for new treatments for this vulnerable pediatric patient population. We hope to demonstrate that dasiglucagon is effective in the prevention of persistent and dangerously low blood sugar levels in these children, which they and their families must live with every day."

In the Phase 3 trial, Zealand will evaluate the potential of long-term dasiglucagon infusion to prevent hypoglycemia in the CHI children. The two-arm, open-label trial comprises up to 32 children with CHI, aged 3 months up to 12 years, and will compare children treated with the current standard of care versus children on the same standard of care plus dasiglucagon infusion. The primary endpoint is the number of hypoglycemic events. The trial is being conducted at sites in the United States, Europe and Israel. Trial details are available at clinicaltrials.gov.

Dr. Indi Banerjee, the Head of the North England CHI NHS service and lead Principal Investigator in the trial said, "I am excited to witness the initiation of the first Phase 3 drug development program specific to this complex rare disease. If successful, it could have significant impact on the brain development of the affected children, and allow their families to regain control of their lives."

A second Phase 3 trial is anticipated to begin in 2019. The trial will similarly evaluate safety and efficacy of administering dasiglucagon via a pump, but in potentially 12 children (aged 7 days to 1 year).

All children deriving benefit from dasiglucagon treatment by the end of these trials will be offered continuation of treatment in a long-term, open-label extension trial.

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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. In 2017, both the U.S. FDA and the European Commission granted orphan drug designation 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.

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