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CHI ANNOUNCES HIHA GRANT AWARD OF \$70,000.00

Glen Ridge, NJ, May 20, 2019 - Congenital Hyperinsulinism International (CHI) is thrilled to announce the award of \$70,000.00 for a one-year pilot grant to Dr. Amanda Ackermann at the Children's Hospital of Philadelphia. With this grant funding, Dr. Ackermann will create a **Novel Mouse Model to Investigate Pathophysiology of Hyperinsulinism/Hyperammonemia Syndrome (HIHA)**. The understanding that will come from creating and studying this mouse model is needed to develop treatments for high blood ammonia levels, seizures, and neurodevelopmental differences caused by HIHA.

The grant was competitively bid internationally, and Dr. Ackermann's project was selected after a careful review of applications by an expert review panel made up of renowned endocrinologists and family members of patients. The committee carefully evaluated the applications which were all excellent.

"I appreciate the focus and dedication to addressing the neurological effects in patients with HIHA. As a parent with a (now adult) child with HIHA, I am passionate about finding alternative treatments for HIHA. Other therapies are needed, and I strongly feel this researcher is acutely aware of this need." - Dina Tallis, EdD, Parent Reviewer of applications.

"The investigator is highly qualified, well trained and in an excellent position to accomplish this project. In my opinion, this project is very appropriate for pilot funding in that it aims at developing a new animal model for this disease that will then be made available to the entire research community for further study of all aspects of this disease. This will be a very valuable resource whose overall impact will extend far beyond this specific grant proposal" - Benjamin Glaser, MD, Professor Emeritus, Hadassah-Hebrew University, Expert Endocrinologist Reviewer of applications.

HIHA is not only a disease of hypoglycemia. Patients with HIHA also have high blood ammonia levels, seizures, and neurodevelopmental differences that currently are not well-understood and do not have any specific treatments. It has been difficult to study each of these features of HIHA in patients because each one can affect the other features.

This project will create a mouse model of HIHA that has the disease-causing mutation only in specific cell types. The initial use this mouse model us to study the effects of the HIHA mutation in the brain, but additional future studies will also be able to use this mouse model to study effects of the HIHA mutation in other organs including the liver and kidneys.

CHI is the leading HI patient advocate dedicated to supporting children and adults born with HI. Founded in 2005 by parents of children with HI, the 501(c)3 organization works to support research leading to better treatments and a cure for the rare disease. The nonprofit raises awareness of the dangers of prolonged hypoglycemia and supports families living with the disease. U.S. donations to CHI are tax deductible.