



Proteostasis Therapeutics Announces Achievement of Preclinical Milestone in Neurodegenerative Diseases Collaboration With Biogen Idec

Company Receives Multimillion Dollar Payment for a Preclinical Milestone

CAMBRIDGE, MA--(Marketwired - July 30, 2014) - Proteostasis Therapeutics, Inc., a company developing novel therapeutics that regulate protein homeostasis to improve outcomes for patients with orphan and neurodegenerative diseases, today announced that it has successfully completed a key preclinical milestone in the collaboration with Biogen Idec. Proteostasis has received a multimillion dollar milestone payment as a result of this achievement.

"We are very encouraged to reach this milestone early in our collaboration with Biogen Idec," said Markus Haerberlein, Ph.D., Chief Scientific Officer at Proteostasis Therapeutics. "Achieving a target protein reduction through inhibition of Usp14 in a whole animal system is an important biological validation and demonstrates the disease-modifying potential of our inhibitors against a variety of neurodegenerative disorders. We look forward to continuing to work with Biogen Idec to bring this program toward candidate designation."

Neurodegenerative diseases, such as Alzheimer's disease and Parkinson's disease, have elevated levels of aggregation-prone proteins in the brain. Prior research has shown that the inhibition of the deubiquinating enzyme, Usp14, enhances proteasome activity and increases the clearance of such proteins.

"We remain optimistic about Usp14 as a very interesting and potentially important target for neurodegeneration," said Spyros Artavanis-Tsakonas, Ph.D., Chief Scientific Officer at Biogen Idec.

In December 2013, Proteostasis and Biogen Idec entered into a collaboration to research and develop therapeutic candidates based on the inhibition of Usp14. Under the agreement, the companies are conducting preclinical research to identify lead compounds for clinical development. At specified points, Proteostasis may opt in for global co-development and U.S. co-

commercialization rights. The total deal contains payments of up to \$200 million, as well as tiered royalties.

About Proteostasis Therapeutics

Proteostasis Therapeutics is developing disease-modifying therapeutics for cystic fibrosis and neurodegenerative diseases. The Company's technology combines both phenotypic and target based drug discovery to develop therapeutics that modulate protein homeostasis pathways and correct for imbalances in protein folding, trafficking, and clearance. For more information, please visit www.proteostasis.com.

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