Proteostasis Therapeutics Announces Expansion of Collaboration with The Scripps Research Institute to Broaden Cystic Fibrosis Program

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Cambridge, Mass., July 26, 2012 — Proteostasis Therapeutics, Inc., a company developing novel therapeutics that regulate protein homeostasis to improve outcomes for patients with neurodegenerative and orphan diseases, announced today that it has expanded its collaboration with The Scripps Research Institute (TSRI) to encompass an additional funded research project focused on biology and the testing of small molecule modulators of protein folding and trafficking for the treatment of cystic fibrosis (CF). This expansion follows the Company's recently announced collaboration with the Cystic Fibrosis Foundation to research, develop, and commercialize therapies to treat patients with the most common mutation of the cystic fibrosis transmembrane conductance regulator (CFTR), Delta F508.

"This expanded collaboration will allow us to accelerate our CF program as we work to advance our most promising series to lead optimization this year," said Mark Enyedy, Chief Executive Officer of Proteostasis Therapeutics.

Working with the laboratory of William Balch, Ph.D., Professor of Cell Biology at TSRI, scientists at Proteostasis Therapeutics have used an integrated platform comprised of genomics, proteomics, functional assays, and medicinal chemistry to identify compounds that regulate key folding and trafficking pathways in the cell. To date, these compounds have demonstrated significant efficacy in CF-specific cellular models. Under the expanded collaboration, Proteostasis Therapeutics will provide funding for this research and will have exclusive rights to license any technology originating from the research.

"We are excited to deepen our relationship with Proteostasis Therapeutics to develop novel approaches that manage the root cause of the problem of this devastating disease," added Dr. Balch.

This newly expanded collaboration will enhance the ability of Proteostasis Therapeutics to perform chaperone-based high throughput screening in multiple disease relevant cellular models to identify Proteostasis Regulators that will correct the folding, trafficking and function of Delta F508 CFTR, both alone and in combination with agents currently in development or on the market.

"Dr. Balch's expertise in CF biology and protein homeostasis complements our proprietary technology for characterizing proteostasis network pathways in normal and disease states. The expansion of this collaboration further underscores our commitment to working with leading academic scientists and institutions in our focus areas in neurodegenerative and orphan diseases," stated Peter Reinhart, Chief Scientific Officer of Proteostasis Therapeutics.

About The Scripps Research Institute

The Scripps Research Institute is one of the world's largest independent, not-for-profit organizations focusing on research in the biomedical sciences. Over the past decades, Scripps Research has developed a lengthy track record of major contributions to science and health, including laying the foundation for new treatments for cancer, rheumatoid arthritis, hemophilia, and other diseases. The institute employs about 3,000 people on its campuses in La Jolla, CA, and Jupiter, FL, where its renowned scientists—including three Nobel laureates—work toward their next discoveries. The institute's graduate program, which awards Ph.D. degrees in biology and chemistry, ranks among the top ten of its kind in the nation. For more information, see www.scripps.edu.

About Proteostasis Therapeutics

Proteostasis Therapeutics is developing disease-modifying therapeutics for orphan and neurodegenerative diseases. The Company's lead programs in cystic fibrosis and Parkinson's disease modulate protein chaperone and proteasomal degradation pathways within the cell. These pathways are part of the cellular 'quality control' machinery, called the

protein homeostasis network or Proteostasis Network (PN) that regulates protein folding, trafficking, and clearance. By enhancing the function and capacity of the Proteostasis Network, the Company's product candidates correct for imbalances in the Proteostasis Network resulting from the cumulative effects of disease, genetic mutations, environmental factors, and aging. For more information, please visit www.proteostasis.com.

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