Proteostasis Therapeutics Announces Collaboration with Cystic Fibrosis Foundation Therapeutics

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– Company to receive funding for novel therapies targeting Delta F508, most common CF mutation –

Cambridge, Mass., May 7, 2012 — Proteostasis Therapeutics, Inc. (PTI), a company developing novel therapeutics that regulate protein homeostasis to improve outcomes for patients with neurodegenerative and orphan diseases, announced today that it has entered into a collaboration with Cystic Fibrosis Foundation Therapeutics, Inc. (CFFT), the non-profit drug discovery and development affiliate of the Cystic Fibrosis Foundation, to research, develop and commercialize therapies to treat patients with the most common cystic fibrosis mutation, Delta F508.

"PTI has a unique technology to help identify new compounds to treat the most common defect in cystic fibrosis and this new collaboration is part of our strategy to find effective therapies for all people with this devastating disease," said Robert J. Beall, Ph.D., President and Chief Executive Officer of the Cystic Fibrosis Foundation.

This collaboration will focus on identifying small molecule modulators of protein homeostasis that will correct the folding, trafficking and function of Delta F508 CFTR. Recent research demonstrates that CF is a disease characterized by improper folding and inadequate trafficking of the cystic fibrosis transmembrane conductance regulator (CFTR) protein. Working closely with collaborators from the laboratory of Dr. William Balch of The Scripps Research Institute (TRSI), PTI scientists 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. These Proteostasis Regulators have demonstrated significant efficacy in CF-specific cellular models. Today's announced
CFFT collaboration will build upon these observations to discover and develop Proteostasis Regulators that correct Delta F508 CFTR function both alone and in combination with agents currently in development and will also expand the company's existing CF biology relationship with TSRI.

"Despite recent progress, improving outcomes for patients with CF remains a significant unmet need, particularly those with the Delta F508 CFTR protein mutation, which is present in over 70 percent of patients," said Mark Enyedy, Chief Executive Officer of PTI. "CFFT has funded some of the most innovative and successful research in the field and we believe this collaboration validates PTI's potential to develop and commercialize novel disease-modifying therapies that will complement existing approaches to managing this devastating disease."

About The Cystic Fibrosis Foundation

The Cystic Fibrosis Foundation is the world's leader in the search for a cure for cystic fibrosis. The Foundation funds more CF research than any other organization, and nearly every CF drug available today was made possible because of Foundation support. Based in Bethesda, Md., the Foundation also supports and accredits a national care center network that has been recognized by the National Institutes of Health as a model of care for a chronic disease. The CF Foundation is a donor-supported nonprofit organization. For more information, go to www.cff.org.

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 (PTI) is developing novel disease-modifying therapeutics that target the cellular pathways regulating protein folding, trafficking, and clearance. The Proteostasis Network (PN) is comprised of these interconnected cellular pathways and can become imbalanced by the cumulative effects of aging, disease, genetics and environmental factors. The Company is focused on improving outcomes for patients with neurodegenerative and orphan diseases with lead programs in cystic fibrosis and Parkinson's disease. For more information, please visit www.proteostasis.com.

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