



Proteostasis Therapeutics and Astellas Announce Collaboration for Research, Development and Commercialization of Therapies Modulating the Unfolded Protein Response

CAMBRIDGE, MA. and TOKYO, JAPAN--(Marketwired - November 04, 2014) - Proteostasis Therapeutics, Inc. ("Proteostasis Therapeutics"), a company developing novel therapeutics to address diseases caused by defects in protein folding, trafficking and clearance, and Astellas Pharma Inc. ("Astellas") today announced that they have entered into a worldwide collaboration to research and develop therapeutic candidates that modulate the Unfolded Protein Response (UPR) through the use of Proteostasis Therapeutics' proprietary "Disease Relevant Translation" (DRT™) and "Proteostasis Network" platform. Proteostasis Therapeutics will receive an initial upfront payment from Astellas, along with a securities investment. Proteostasis Therapeutics is also eligible for research funding support, future development and commercial milestones that could result in total payments of over \$400 million, as well as tiered royalties. Further, Astellas has the right to begin two additional projects under the same terms, which, if it fully exercised this right, would bring the total potential value of the collaboration to \$1.2B.

"We are very pleased to be collaborating with Astellas, a worldwide leader in the development of innovative therapeutics," said Meenu Chhabra, President and CEO at Proteostasis Therapeutics. "Our novel approach to drug discovery, coupled with Astellas' track record in drug development, will enable rapid discovery and development of therapies for important unmet medical needs."

The collaboration will focus on one genetic disease, and further explore additional indications that can be affected through modulation of the UPR pathway. Stress induced by accumulation of unfolded proteins in the endoplasmic reticulum (ER) is observed in many diseases which are now recognized as protein conformational diseases, including genetic diseases, neurodegenerative diseases, and retinal degenerative diseases. Selective modulation of the UPR pathway in non-clinical investigations improved the stress response and restored function, suggesting that it can be beneficial as potential novel disease-modifying therapies for

multiple diseases with high unmet medical needs.

"Proteostasis Therapeutics has a novel platform that offers a differentiated approach to discovering drugs for unmet medical needs," commented Kenji Yasukawa, Ph.D., Senior Vice President and Chief Strategy Officer, at Astellas, "it complements our existing internal and externalized R&D initiatives and we continue to invest in innovative technologies that can provide new therapeutic options to patients. We look forward to working closely with Proteostasis Therapeutics to identify lead candidates for clinical development and potential commercialization."

As part of the agreement, the Companies will conduct discovery, screening and preclinical research to identify lead compounds for clinical development. Upon candidate selection, Proteostasis Therapeutics will have the rights to opt in for global co-development and United States co-promotion.

This collaboration is being led by Innovation Management ("AIM") of Astellas Pharma Inc.

The impact of this collaboration has been accounted in revised Astellas' forecasts for fiscal year ending March 2015, which was announced on October 31st, 2014.

About Proteostasis Therapeutics, Inc.

Proteostasis Therapeutics, Inc. is a drug discovery company addressing diseases caused by defects in protein folding, trafficking and clearance for orphan and neurodegenerative diseases. The Company's Disease Relevant Translation (DRT™) platform combines a proprietary screening approach with state-of-the-art medicinal chemistry, to generate highly selective drug candidates and is advancing a pipeline inclusive of a lead program in cystic fibrosis and a partnership with Biogen Idec for neurodegenerative diseases.

The Company's DRT™ platform utilizes functionally pertinent assays and disease relevant models to identify highly translatable therapies associated with the modulation of protein homeostasis pathways within the cell. These pathways are part of cellular 'quality control' machinery, known as the protein homeostasis network or Proteostasis Network (PN). By modifying the function and capacity of the PN, the Company's therapeutic product candidates correct for imbalances in the PN resulting from the cumulative effects of disease, genetic mutations, environmental factors and aging. For more information visit www.proteostasis.com.

About Astellas Pharma Inc.

Astellas Pharma Inc., located in Tokyo, Japan, is a pharmaceutical company dedicated to improving the health of people around the world through the provision of innovative and reliable pharmaceuticals. Astellas has approximately 18,000 employees worldwide. The organization is committed to becoming a global category leader in Urology, Immunology (including Transplantation) and Infectious diseases, Oncology, Neuroscience and Diabetes Mellitus (DM) Complications and Kidney diseases. For more information on Astellas Pharma Inc., please visit the company website at www.astellas.com/en.

About Innovation Management

Innovation Management ("AIM") is a new division established in October 2013 to enhance and accelerate the process of screening and realizing external opportunities to strengthen innovation at the preclinical development stage. AIM oversees strategic alliance activities with external partners and is responsible for a series of activities at acquiring external innovation opportunities in the preclinical development stage, such as strategy planning, screening, scientific assessment and alliance negotiations, so that strategic external business alliances

can be strategically and systematically performed.

Investor Contacts:

Susan Shepard
Proteostasis Therapeutics, Inc.
sshepard@proteostasis.com
(617) 225-0096 x2113

Malini Chatterjee
Blueprint Life Science Group
mchatterjee@bplifescience.com
(917) 330-4269

Astellas Contacts:

Astellas Pharma Inc.
Corporate Communications
+81-3-3244-3201