



LECTURE

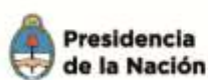
ER proteostasis disturbances in neurodegenerative diseases.

Dr. CLAUDIO HETZ

He was originally trained as Biotechnology Engineer at the University of Chile and performed a Ph.D in Biomedical Sciences at Serono Pharmaceutical Research Institute, Switzerland. Then he did his postdoctoral training at Harvard University. He joined the University of Chile during 2007 and is currently Full Professor at Faculty of Medicine and adjunct Professor at Harvard. He is also currently the Co-Director of the Biomedical Neuroscience Institute. He has received important award including the TWAS-ROLAC Young Scientist Prize as outstanding young scientist in Latin America, was finalist in the Eppendorf and Science Award in Neurobiology, and was awarded with the Cell Biology Society and Bios-Chile prize as the best young scientist of Chile. Recently he obtained the KIA international award for his contributions to biomedicine.

Our laboratory focuses on understanding the molecular basis of organelle stress and its relationship to pathological conditions affecting the nervous system, and the development of prototypic therapies to prevent this damage. Our laboratory is committed to the study of cellular strategies involved in adaptation to chronic endoplasmic reticulum (ER) stress. The ER has important cellular functions, highlighting its role as sophisticated machinery for protein folding and secretion. ER stress engages an integrated signaling pathway known as the “Unfolded Protein Response” (UPR), which aims to restore homeostasis. Nevertheless, the mechanisms that control the transition from an adaptive state to cell death processes remain unknown and is a central subject of our research. We are currently developing a systematic approach to underscore the effects of targeting the UPR in several brain diseases involving protein misfolding using genetic manipulation of the pathway in mouse models in addition to develop gene therapy strategies to alleviate ER stress. Our laboratory is one of the most productive groups in Chile and supported by various national and international organizations. Dr Hetz has published more than 100 paper with more than 5200 citations (H-Factor 35).

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