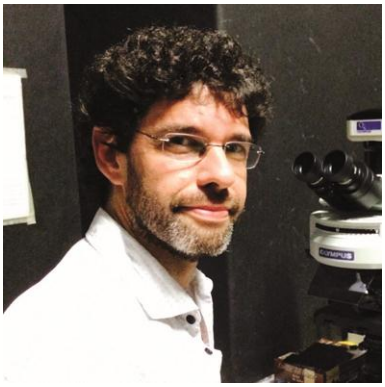

LECTURE

Conditional mouse models of TDP-43 proteinopathies.

Dr. LIONEL MÜLLER IGAZ

He is a career Investigator (PI) at the National Council for Scientific and Technological Research (CONICET) of Argentina, and Director of the Neuronal Physiopathology Laboratory, Institute Houssay for Physiology and Biophysics (IFIBIO-Houssay) – CONICET, located a the University of Buenos Aires (UBA) School of Medicine (Buenos Aires, Argentina). He graduated as Licenciado (equivalent to a BsC) in Biological Sciences in 2000 (UBA). He obtained his PhD in Neuroscience from the University of Buenos Aires (2005) and was a postdoctoral fellow at the Center for Neurodegenerative Research (CNDR) at the University of Pennsylvania, USA (2006-2010). He is a member of several scientific societies, including International Brain Research Organization (IBRO), Society for Neuroscience (SfN), International Society for Neurochemistry (ISN) and Argentine Society for Neuroscience Research (SAN).

During his scientific career, Lionel Müller Igaz has made contributions to a number of different topics. As an undergrad, he first studied neuro-immunoendocrine interactions during thymocyte selection. During his doctoral studies, he contributed in establishing the existence of different time periods of transcription necessary for memory consolidation in mammals, as well as the role of mRNA and protein synthesis during memory extinction and persistence. As a postdoc at Upenn he studying the role of the protein TDP-43 in neurodegenerative disease initially characterizing the different TDP-43 pathological species found in the patients with TDP-43 proteinopathies such as FTD and ALS, finding regional (brain vs. spinal cord) differences in inclusion composition. Later he focused on developing cellular models that recapitulate the disease process through expression of different TDP-43 species (mainly C-terminal fragments). In parallel, he developed and characterized novel TDP-43 transgenic animal models, including those with inducible, forebrain neuron-enriched expression of either nuclear or cytoplasmic forms of TDP-43.

In his recently established lab at the University of Buenos Aires, Argentina, he continues to explore the pathophysiological roles of TDP-43 in the nervous system. Recently, his group described a thorough behavioral characterization of the transgenic mouse expressing the cytoplasmic form of TDP-43, showing motor, cognitive and social changes that recapitulate those seen in FTD/ALS, and also demonstrating the reversibility of some of these deficits upon transgene suppression. He is currently pursuing different lines of research integrating multidisciplinary approaches (including behavioral, pharmacological, biochemical and molecular studies) to address the pathogenic mechanisms underlying TDP-43 proteinopathies, which in turn will be vital to develop new and more effective therapies for these disorders.