Prusiner-Abramsky Research Awards

Previous Winners 2014

Prof. **ALBERT TARABOULOS**
Department of Microbiology and Molecular Genetics
Institute for Medical Research – Israel-Canada
Hebrew University-Hadassah Medical School
Prion Neurotoxicity: From Protein Misfolding to Lipid Disease

Prof. **HAGAI BERGMAN**
Department of Medical Neurobiology
Institute for Medical Research – Israel-Canada
Hebrew University-Hadassah Medical School
Computational Physiology and Pathophysiology of the Basal Ganglia and their Disorder – From Understanding to Closed Loop Deep Brain Stimulation Treatments

Dr. **DANA EKSTEIN**
Department of Neurology
Hebrew University-Hadassah Medical School
Development of Tools for Patient-Specific Individualized Diagnosis and Treatment of Epilepsy

Dr. **ADI VAKNIN-DEMBINSK**
Department of Neurology
Hebrew University-Hadassah Medical School
Personalized Medicine in Multiple Sclerosis and Neuromyelitis Optica: Predicting Disease Outcome and Treatment Responsiveness

Previous Winners 2013

Prof. **ALEXANDER LOSSOS**
Department of Neurology
Hebrew University-Hadassah Medical School
Diagnosis and Treatment of Adult Polyglucosan Body Disease

Dr. **HANNA ROSENMANN**
Department of Neurology
Hebrew University-Hadassah Medical School
Alzheimer's Disease and Tauopathies - Improved Animal Models, Pathogenesis and Therapeutic Approaches

Dr. **SARA EYAL**
Institute for Drug Research
School of Pharmacy
Imaging CNS Function in Health and Disease

Dr. **ADI INBAL**
Department of Medical Neurobiology
Institute for Medical Research – Israel-Canada
Hebrew University-Hadassah Medical School
Molecular Mechanisms of Forebrain and Eye Development

Previous Winners 2012

Dr. **RONIT SHARON**
Institute for Medical Research Israel-Canada
Hebrew University-Hadassah Medical School

Prof. **RONEN LECKER**
Department of Neurology
Hebrew University-Hadassah Medical School

Dr. **ALEXANDER M. BINSHTOK**
Institute for Medical Research Israel-Canada
Hebrew University-Hadassah Medical School

Prof. **HAIM OVADIA**
Department of Neurology
Hebrew University-Hadassah Medical School

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June 2015

The Authority for Research and Development
The Hebrew University of Jerusalem

Project coordinator: Ayelet Sagiv
Project editor: Shoshana Glatzer
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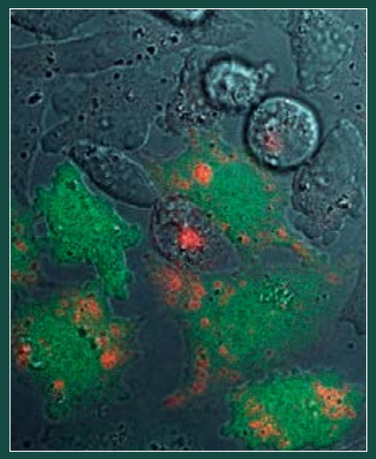
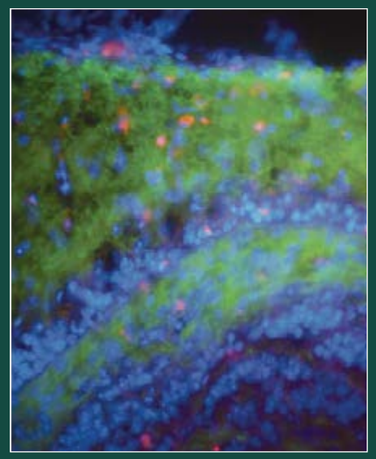
THE PRUSINER-ABRAMSKY RESEARCH AWARDS

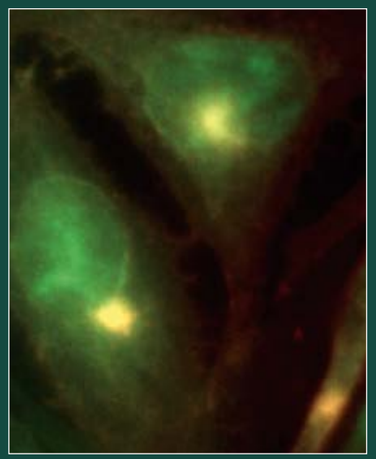
AT THE HEBREW UNIVERSITY OF JERUSALEM

BY THE ORION FOUNDATION

תשע"ה 2015







90 years

האוניברסיטה העברית בירושלים
THE HEBREW UNIVERSITY OF JERUSALEM

PRUSINER-ABRAMSKY

Research Awards - 2015

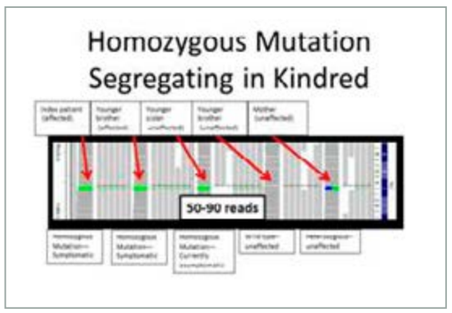
Prof. Stanley B. Prusiner, M.D.

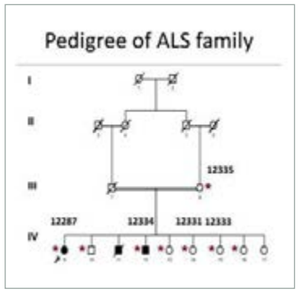
Stanley B. Prusiner, M.D., is Director of the Institute for Neurodegenerative Diseases and Professor of Neurology at the University of California, San Francisco (UCSF), where he has worked since 1972. Born in Des Moines, Iowa, in 1942, he spent his childhood there and in Cincinnati, Ohio. He received his undergraduate degree and medical training at the University of Pennsylvania and his postgraduate clinical training at UCSF. From 1969-72, he served in the U.S. Public Health Service at the National Institutes of Health. He is the author of over 500 research articles and the book *Madness and Memory*.

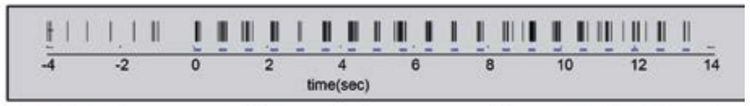
Prof. Prusiner is a member of the U.S. National Academy of Sciences, the Institute of Medicine, the American Academy of Arts and Sciences, the American Philosophical Society, and a foreign member of the Royal Society of London. He is the recipient of numerous prizes, including the Potamkin Prize for Alzheimer's Disease Research of the American Academy of Neurology (1991); the Richard Lounsbery Award for Extraordinary Scientific Research in Biology and Medicine from the National Academy of Sciences (1993); the Gairdner Foundation International Award (1993); the Albert Lasker Award for Basic Medical Research (1994); the Paul Ehrlich Prize from the Federal Republic of Germany (1995); the Wolf Prize in Medicine from the State of Israel (1996); the Keio International Award for Medical Science (1996); the Louisa Gross Horwitz Prize from Columbia University (1997); the Nobel Prize in Physiology or Medicine (1997); and the U.S. National Medal of Science (2009).

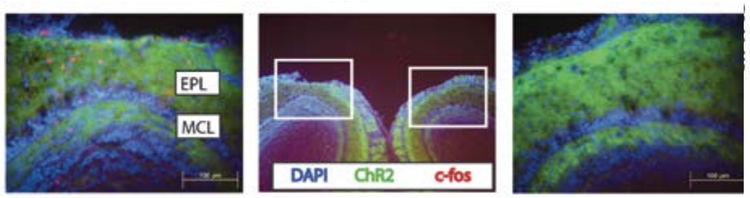
Prof. Prusiner's groundbreaking research on prion diseases, beginning in the late 1970s, led him to propose an explanation for the cause of bovine spongiform encephalopathy ("mad cow" disease) and its human equivalent, Creutzfeldt-Jakob disease, for which he was awarded the Nobel Prize. In this work, he coined the term prion (derived from "proteinaceous" and "infectious") to refer to a previously undescribed form of infection caused by the self-propagation of alternatively folded proteins.

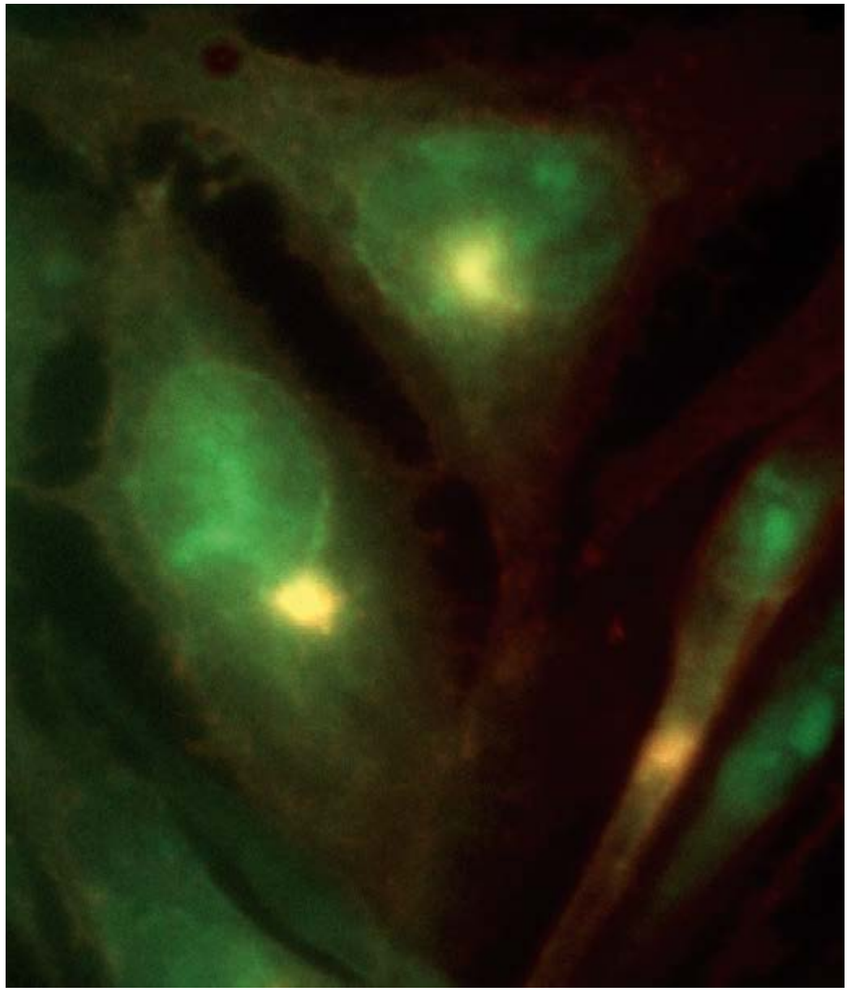
His research has elucidated a fundamental understanding of the proteins underlying such illnesses as Alzheimer's disease, Parkinson's disease, amyotrophic lateral sclerosis (ALS) and PrP prion diseases. These advances in understanding the molecular, genetic and cellular basis of neurodegenerative diseases have fueled progress toward the development of targeted drug therapies.

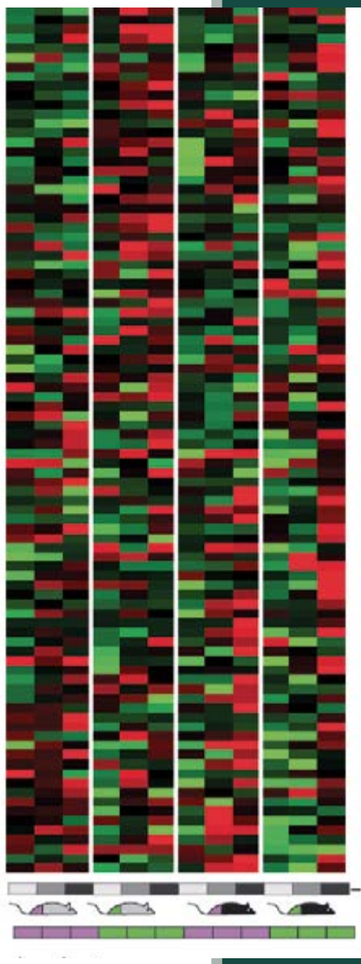












PRUSINER-ABRAMSKY

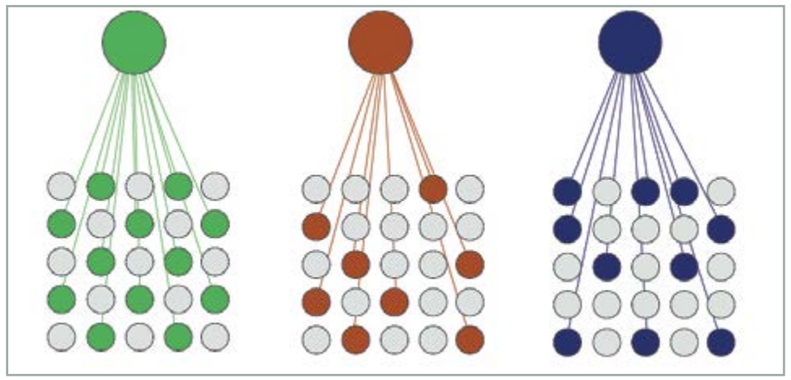
Research Awards - 2015

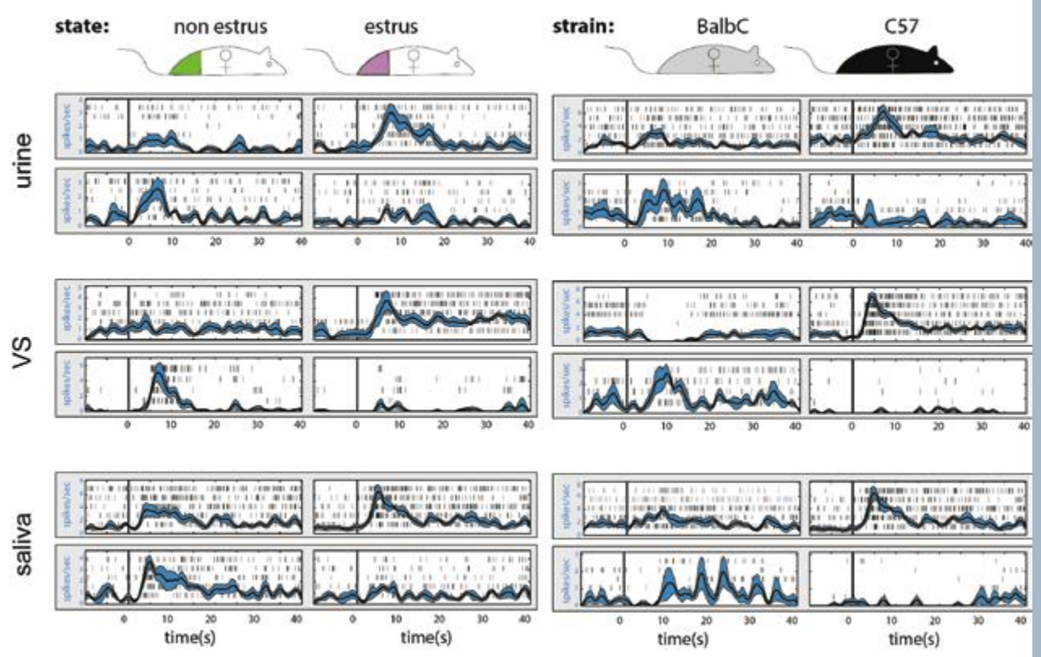
Prof. Oded Abramsky, M.D., Ph.D.

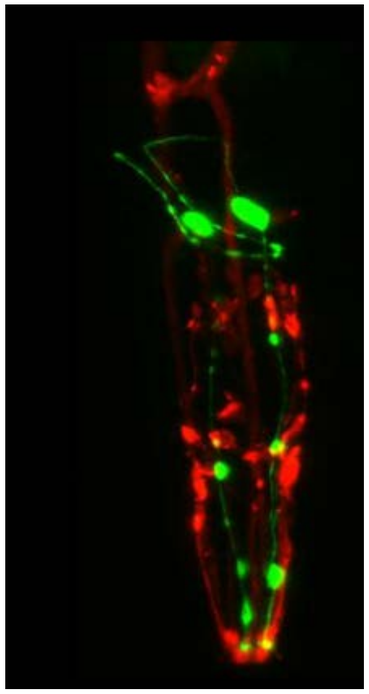
Oded Abramsky was born in Jerusalem and received his M.D. and Ph.D. degrees from The Hebrew University of Jerusalem. He completed his residency in neurology at Hadassah University Hospital, where he was later appointed Head of the Neuroimmunology Unit (1982) and Chairman of the Neurology Department (1988-2005). He was appointed Professor of Neurology at the Hebrew University-Hadassah Medical School in 1982, holding the Israel S. Wechsler Chair in Neurology. He served as Dean of the Faculty of Medicine of the Hebrew University (1992-96) and subsequently was appointed Chairman of the Agnes Ginges Center for Human Neurogenetics at Hadassah University Medical Center.

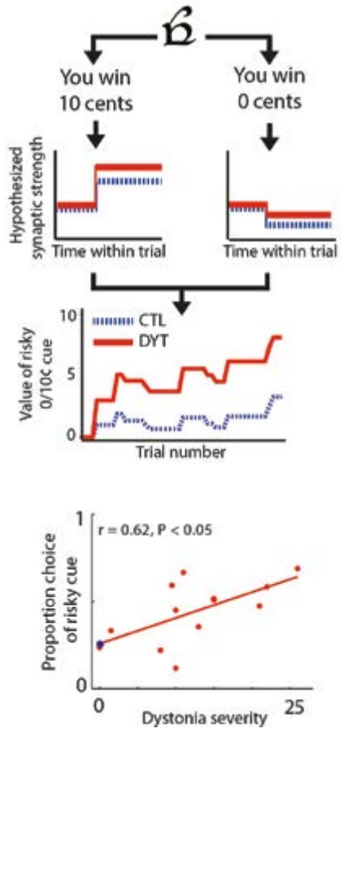
Prof. Abramsky has been actively involved in many aspects of medical research and holds prominent positions in numerous professional organizations concerned with both clinical practice and medical research. He was Chief Scientist of the Israel Ministry of Health (1987-1992), Chairman of the National Medical Research Organization, and served as Chairman of the Israel National Council for Research and Development. He is an Honorary President of the Israel Society of Neuroimmunology; Honorary Member of the American Neurological Association; Member of the Institute of Medicine, National Academy of Sciences (USA); Fellow by Distinction of the Royal College of Physicians (FRCP); and Member of the Israel Academy of Sciences and Humanities, among many other affiliations. In 2008, the Oded Abramsky Chair in Neuroimmunology was established in his honor by Biogen USA at the Hadassah University Medical Center.

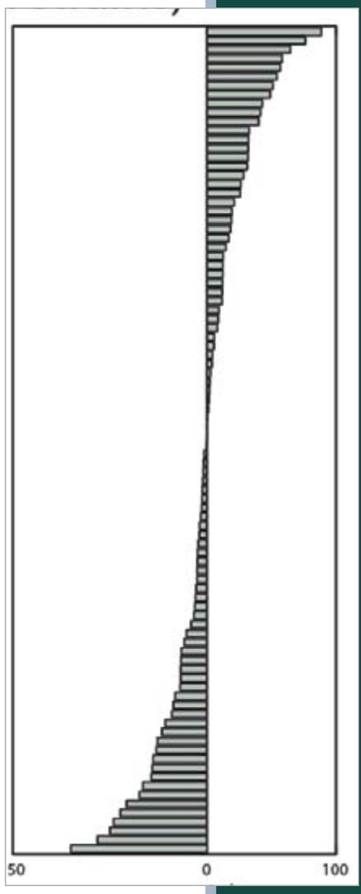
Prof. Abramsky's clinical and scientific research focuses on autoimmune neurological diseases. He was a pioneer in the field of neuroimmunology and demonstrated immune pathogenesis in various neurological diseases of the central and peripheral nervous systems and muscle. Indeed, he proved that myasthenia gravis (MG) is an autoimmune disease, and showed the beneficial effect of corticosteroids and chemotherapy on induced experimental MG. His research served as a guideline to successful immunotherapy of MG and many other autoimmune diseases.











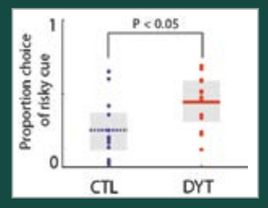
PRUSINER-ABRAMSKY

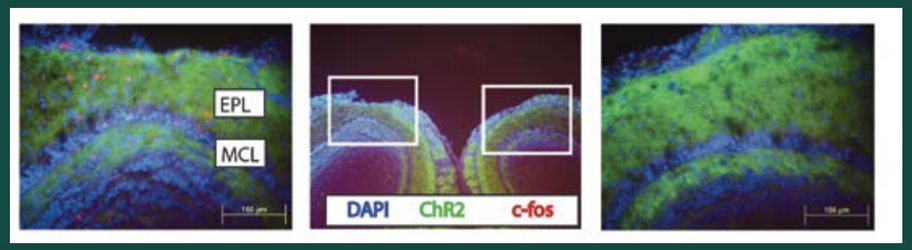
Research Awards - 2015

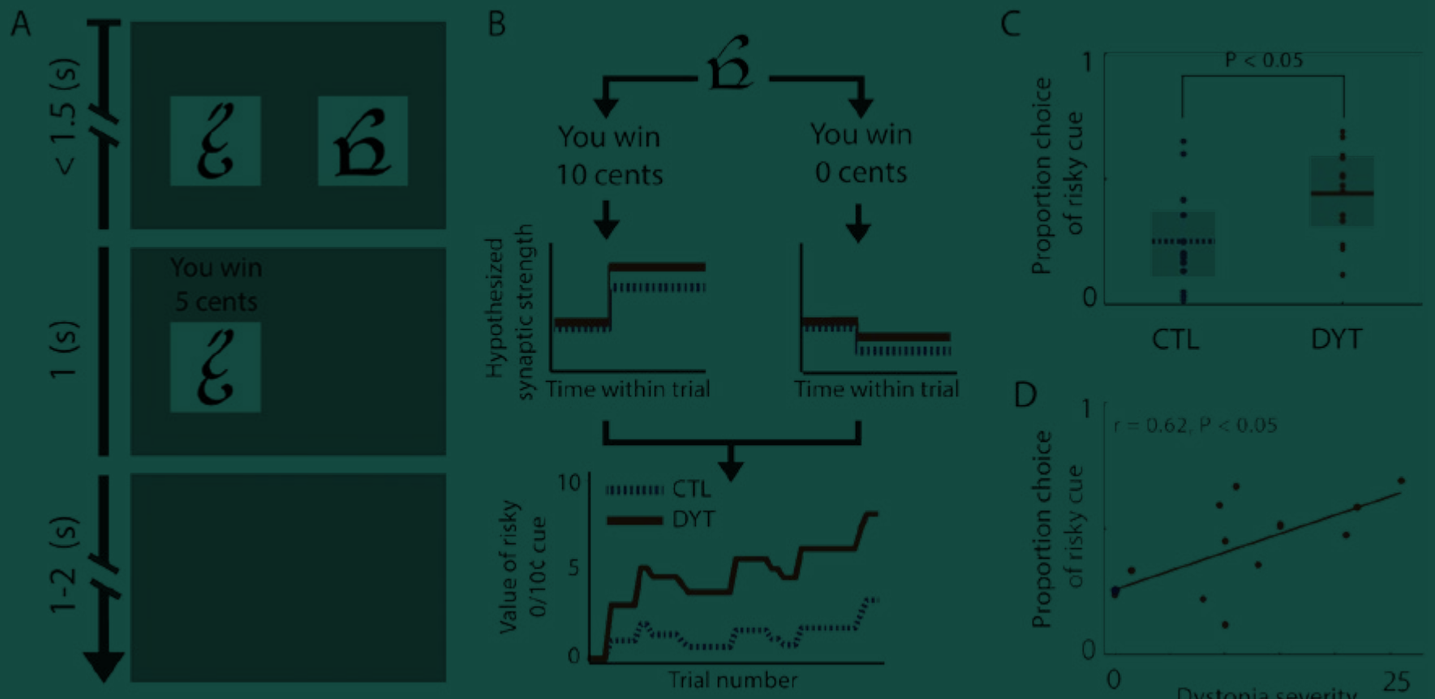
The prestigious **Prusiner-Abramsky Research Awards in Clinical and Basic Neuroscience by The Orion Foundation** honor Professors Stanley Prusiner and Oded Abramsky. Prof. Prusiner of the University of California at San Francisco is a Nobel Prize Laureate in Medicine (1997) and an Honorary Doctor of the Hebrew University of Jerusalem.

Prof. Abramsky is the former Chairman of the Neurology Department and a former Dean of the Faculty of Medicine at Hebrew University.

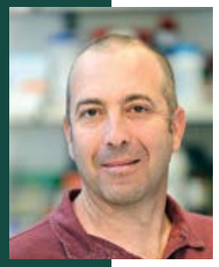
The awards are intended for outstanding researchers from all fields of basic clinical neurosciences at the Hebrew University and the Hadassah University Medical Center.







RESEARCHERS

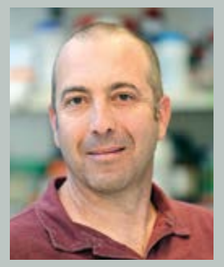


Dr. EHUD COHEN
Department of Biochemistry and Molecular Biology
Institute for Medical Research Israel-Canada
Hebrew University-Hadassah Medical School
Faculty of Medicine
Dissecting the Mechanistic Roles of Aging
in the Emergence of Neurodegenerative Disorders

Dr. YORAM BEN-SHAUL
Department of Medical Neurobiology
Institute for Medical Research Israel-Canada
Hebrew University-Hadassah Medical School
Faculty of Medicine
Neuronal Circuits Underlying Social Behavior

Dr. MARC GOTKINE
Department of Neurology
Hebrew University-Hadassah Medical School
Faculty of Medicine
Identification of Serological, Cytological and Genetic Factors
Associated with the Development and Progression of ALS in Israel

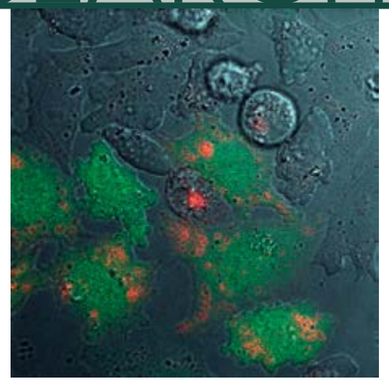
Dr. DAVID ARKADIR
Department of Neurology
Hebrew University-Hadassah Medical School
Faculty of Medicine
DYT1 Dystonia Links Corticostriatal Synaptic Plasticity and Learning
Behavior in Humans

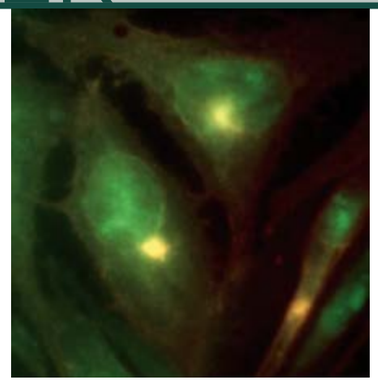


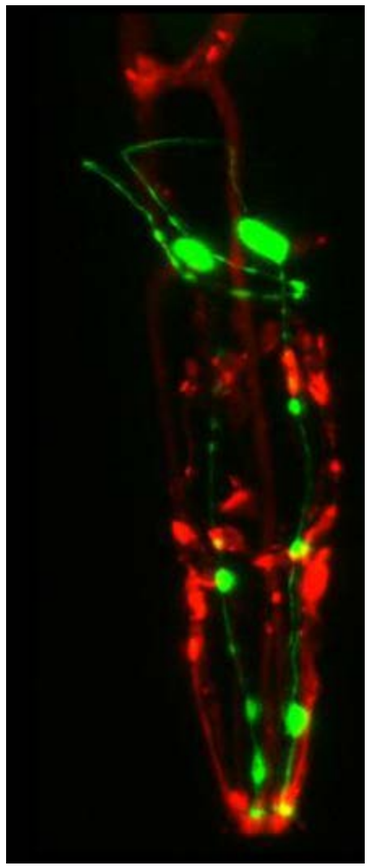
Dr. Ehud Cohen
Department of Biochemistry and Molecular Biology, Institute for Medical Research Israel-Canada
Hebrew University-Hadassah Medical School

As a Ph.D. student at the School of Medicine of the Hebrew University, Ehud Cohen studied the cell biological mechanisms that underlie the development of late-onset familial prion disorders. Why these maladies manifest late in life and whether the manipulation of aging can protect against these illnesses were the questions that he focused on as a post-doctoral fellow at the Salk Institute for Biological Studies (La Jolla, California). In 2009, Dr. Cohen joined the faculty of the Hebrew University. In his laboratory, the cell biology and genetics of aging are investigated as well as the links between this process and neurodegenerative disorders.

RESEARCHER







Dissecting the Mechanistic Roles of Aging in the Emergence of Neurodegenerative Disorders

Aging manipulation is an emerging strategy aimed to postpone the manifestation of late-onset neurodegenerative disorders such as Alzheimer's (AD) and Huntington's diseases (HD) and to slow their progression once emerged. Previously, we found that the alteration of aging by reducing the activity of the Insulin/IGF signaling cascade (IIS), a prominent aging regulating pathway, protects worms from proteotoxicity of various aggregative proteins, including the AD-associated peptide, Aβ. Similarly, IGF1 signaling reduction protects mice from AD-like disease. These discoveries suggest that IIS inhibitors can serve as new drugs for the treatment of neurodegenerative maladies including AD. Our research focuses on the mechanistic links between the aging process and protein quality control mechanisms, asking why neurodegenerative maladies onset late in life. Specifically, we study protein aggregation and deposition at the cellular level, the effects of inter-tissue communication on protein homeostasis (proteostasis) in nematodes, and the therapeutic potential of IGF signaling inhibitors in mice. Our overall goal is to develop efficient counter-neurodegeneration therapies by selective manipulation of aging.

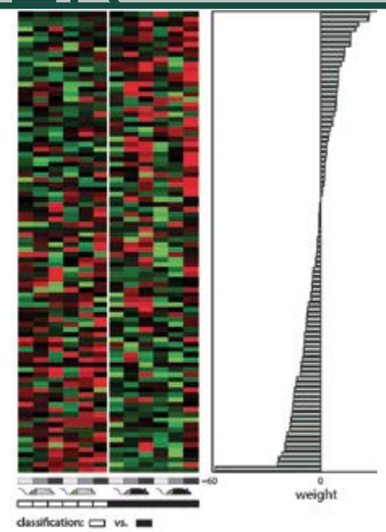
Dr. Yoram Ben-Shaul
Department of Medical Neurobiology, Institute for Medical Research Israel-Canada
Hebrew University-Hadassah Medical School

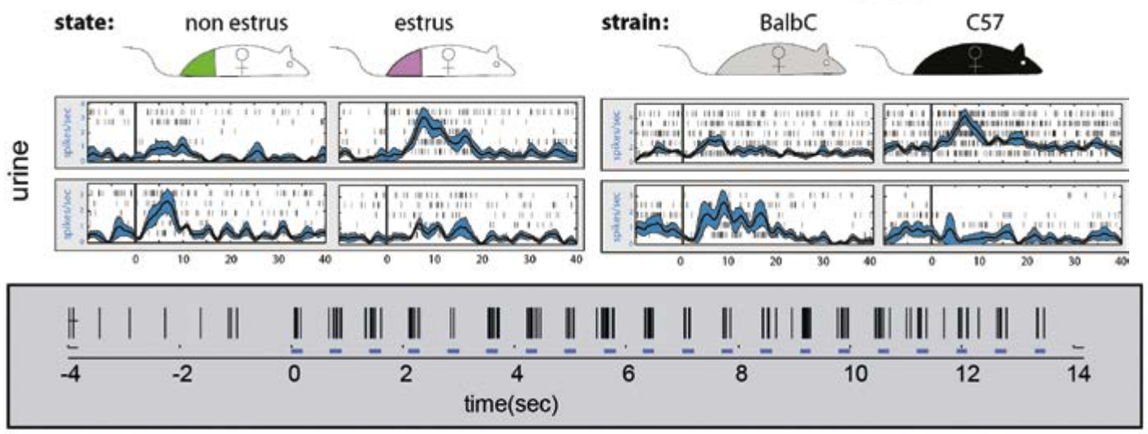
Yoram Ben-Shaul completed his Ph.D. (summa cum laude) with Prof. Moshe Abeles at Hebrew University's Interdisciplinary Center for Neural Computation, using electrophysiology to study encoding of complex movements by the primate motor cortex. As a post-doc with the late Prof. Larry Katz at Duke University, he studied how social information is represented by the vomeronasal system. Following Prof. Katz's untimely death, he continued his postdoctoral work with Prof. Catherine Dulac at Harvard, where his research shifted to studying how central brain regions process social information. He returned to Israel in 2011 with his wife and 3 children, joining the faculty of Hebrew University as a senior lecturer.

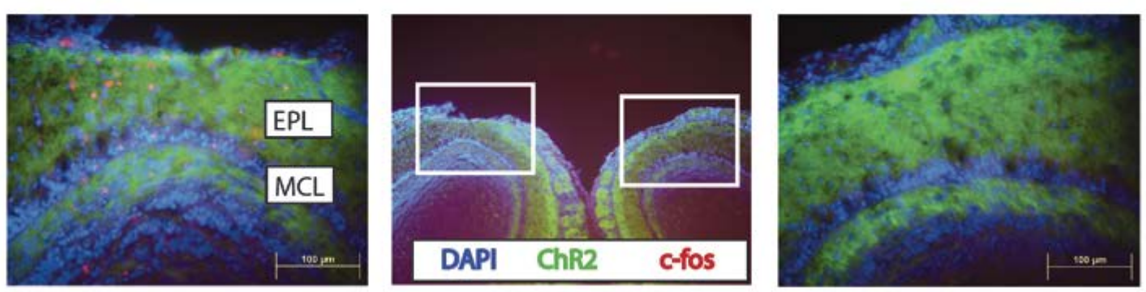
RESEARCHER

Neuronal Circuits Underlying Social Behavior

Dr. Ben-Shaul's research aims to understand how socially relevant information is derived from chemical cues, particularly by the vomeronasal system. His research team mainly uses in-vivo electrophysiology, but also optogenetics and behavior, allowing them to measure neuronal activity, and also to manipulate it and evaluate its consequences. Using these approaches, their research is organized around several topics: neuronal representations of social features, influence of an organism's own physiological state on information processing, neuronal mechanisms underlying social memories, structure function relationships in the vomeronasal system, and, distinctions between innate and learned behaviors.



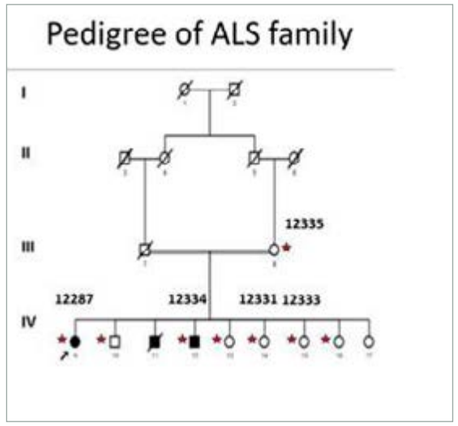




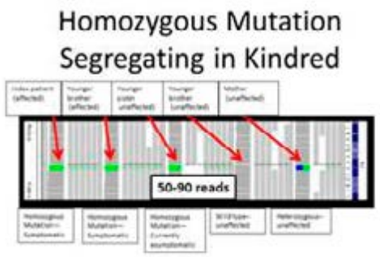
Dr. Marc Gotkine
Department of Neurology
Hebrew University-Hadassah Medical School

Marc Gotkine studies the genetics and epidemiology of ALS, and ALS biomarkers. After receiving his medical degree from King's College, London, he immigrated to Israel where he completed his residency in the Department of Neurology at Hadassah University Medical Center. He is currently in charge of the neuromuscular service and established the Hadassah ALS Multidisciplinary Clinic in 2008, where he created a dedicated ALS database application for data collection from ALS patients. This enabled recognition of phenotypic disease subtypes, potential environmental risk-factors and putative biomarkers which can be correlated with detailed clinical data from the database. Findings indicated that a number of ALS patients were from consanguineous families. He hypothesized that genetic analysis of this specific patient subgroup could lead to the discovery of new pathogenic mutations causing ALS.

RESEARCHER



Identification of Serological, Cytological and Genetic Factors Associated with the Development and Progression of ALS in Israel



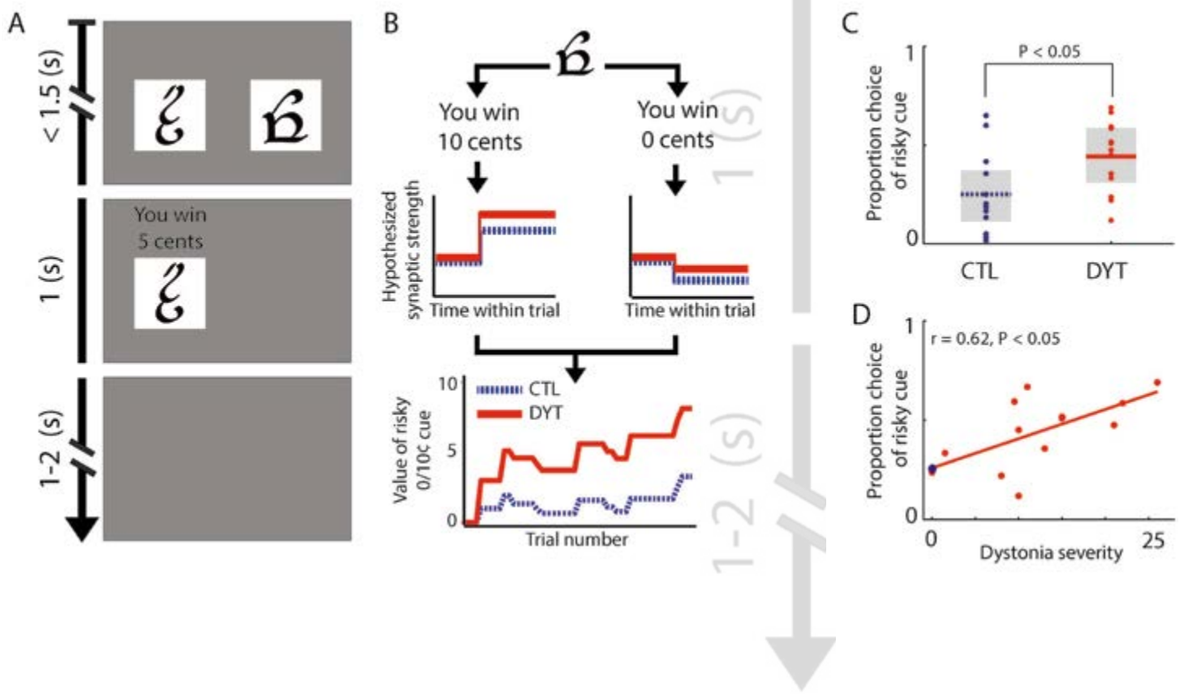
The distinctive properties of the Israeli population—large families with high levels of consanguinity—together with our observation that many ALS patients were from consanguineous families, led us to hypothesize that some of these patients were manifesting pathogenic recessive mutations. To investigate this, patients confirmed to have ALS were asked specifically about consanguinity; when confirmed, they were consented for DNA analysis and interviewed regarding potentially related conditions in family members. I recently examined a Palestinian woman—the product of a consanguineous relationship (her parents were first-cousins)—with an aggressive form of ALS; there was no history of neurodegenerative diseases in previous generations, however examination of medical records confirmed the presence of ALS (previously undiagnosed) in two additional siblings (Figure 1: Pedigree of the kindred). After common mutations were excluded, exome sequencing of the index patient and her siblings (in collaboration with Professor Christopher Shaw's group in London) revealed a novel homozygous mutation, predicted to cause complete loss of function of a protein known to be critical to motor neuron integrity (Figure 2: homozygous mutation). We have scheduled skin biopsies for generating induced pluripotent stem cells, which I will investigate in Prof. Shaw's laboratory in London, in order to prove the pathogenicity of the mutation.

Dr. David Arkadir
Department of Neurology
Hebrew University-Hadassah Medical School

During the last two years, David Arkadir focused his research on the subject of learning processes in patients with DYT1 dystonia, aiming to shed light on the role of corticostriatal plasticity in generating this disabling movement disorder. He developed an interest in movement disorders, including its research and clinical aspects, while training in neurology. at the Hebrew University-Hadassah Medical School under the supervision of Prof. Tamir Ben-Hur. He also recently completed a fellowship in Movement Disorders at Columbia University Medical Center, under the supervision of Dr. Stanley Fahn. His Ph.D. research, conducted in the laboratory of Prof. Hagai Bergman at Hebrew University, focused on the characteristics of information flow in the basal ganglia, a topic that is particularly relevant to understanding of the pathophysiology of movement disorders.

RESEARCHER

DYT1 Dystonia Links Corticostriatal Synaptic Plasticity and Learning Behavior in Humans



It has been difficult to link synaptic modification to overt behavioral changes. Rodent models of DYT1 dystonia, a monogenetic movement disorder, demonstrate increased long-term potentiation and decreased long-term depression in corticostriatal synapses. Computationally, such asymmetric learning predicts risk taking in probabilistic tasks. Using non-invasive behavioral task and functional MRI we learn abnormal risk-taking in DYT1 dystonia patients. This project aims to reveal the role of striatal plasticity in shaping both the motor manifestations of dystonia and choice behavior in humans.