DMM launches new special collection on neurodegeneration

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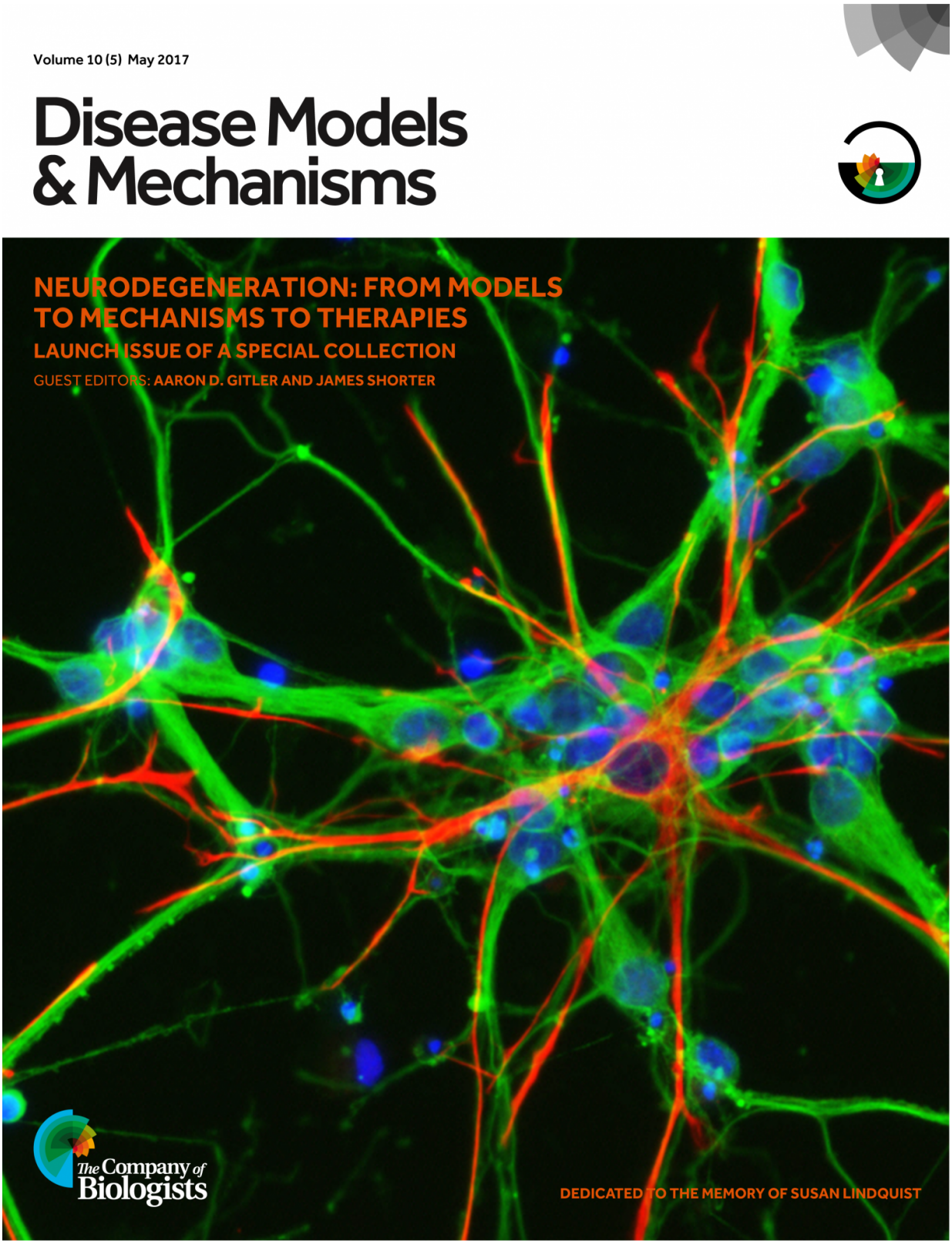

The cover of the DMM special issue showing primary cortical neuronal culture

isolated from newborn mice. Credit: Vito Antonio Baldassarro

A growing aging population means that age-dependent neurodegenerative disorders such as Alzheimer's disease, Parkinson's disease, Huntington's disease and amyotrophic lateral sclerosis are affecting increasing numbers of people, making research into these diseases more important than ever. As a crucial resource for researchers and to showcase the vital research being done in the field, *Disease Models & Mechanisms* (DMM) is launching a new special collection - Neurodegeneration: from models to mechanisms to therapies.

A special issue of DMM is being published to launch the collection, guest edited by Aaron Gitler (Stanford University) and James Shorter (the Perelman School of Medicine at the University of Pennsylvania), both renowned specialists in protein-misfolding neurodegenerative disorders. Our guest editors open the issue with an editorial outlining historical models in neurodegeneration research, and the potential for future breakthroughs using cutting-edge induced pluripotent stem cell (iPSC) and 3D brain organoid models. They also discuss new therapies such as the recent successful clinical trials for antisense oligonucleotide (AO) therapy for spinal muscular atrophy, outline articles in the special issue and highlight key neurodegeneration articles published previously in DMM.

Following the Editorial, DMM Founding Editor Huda Zoghbi describes in an interview how the cases she saw as a clinical neurologist inspired her to move into the lab. A long-time advocate of using model systems in human disease research and epitomising the dedication and passion of the researchers in the field, Huda shares why it has taken her 25 years to understand the biology of neurological disorders well enough to start developing therapeutics. Another highlight of the special issue is a poster

by Edward Lee and colleagues providing a detailed visual realisation of how impaired RNA metabolism can underlie the pathophysiology of neurodegeneration, as well as discussing potential therapeutic interventions to target these processes.

Spotlight on neurodegenerative disorders

Amyotrophic lateral sclerosis (ALS) is covered extensively in this special issue, beginning with a review by Wim Robberecht and colleagues, which provides a comprehensive discussion of model systems used to study ALS, from cell-based systems to fruit flies to rodents. A research article from Javier Fernández-Ruiz and colleagues describes a new spontaneous canine degenerative myelopathy model for ALS that offers several advantages over traditional models relying on transgenic overexpression. Further research articles discuss the molecular basis for weight loss in neurodegenerative disorders with a focus on tyrosine receptor kinase 3 (Kim et al., 2017), the role of autophagy dysregulation in ALS (Wald-Altman et al., 2017), and a new Ranbp2 knockout mouse model for studying the impairments in nucleocytoplasmic transport that can underlie neurodegenerative diseases (Cho et al., 2017).

A second review article presents a discussion by Gabriela Caraveo and colleagues on the current evidence for dysregulation of calcium-dependent pathways in Parkinson's disease (PD), and its implications in drug development. Research articles focusing on PD describe how an analysis of blood samples from a large Turkish family revealed complexin-1 as potential new blood biomarker in early PD risk screening (Lahut et al., 2017), and how genetic mutations linked to PD lead to stage-specific deregulation of the nucleolus (Evsyukov et al., 2017).

Less well-known neurodegenerative disorders also feature in this special issue, including an in vitro cellular model exploring Miller-Fisher syndrome (Rodella et al., 2017), and a study into the role of the

IKBKAP gene in hereditary sensory and autonomic neuropathies (HSANs) of the peripheral nervous system (Chaverra et al., 2017).

Remembering Susan Lindquist

No compilation of current neurodegeneration research would be complete without taking a moment to reflect on the extraordinary contributions to the field by Susan Lindquist, who passed away last year. Her legacy is clear in the number of articles in DMM authored by, citing or mentioning Susan, including a poster co-authored with Julie Valastyan that provides a snapshot of protein-level mechanisms implicated in neurodegenerative and related disorders, and a moving tribute by Vivian Siegel. This special collection is dedicated to her memory.

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