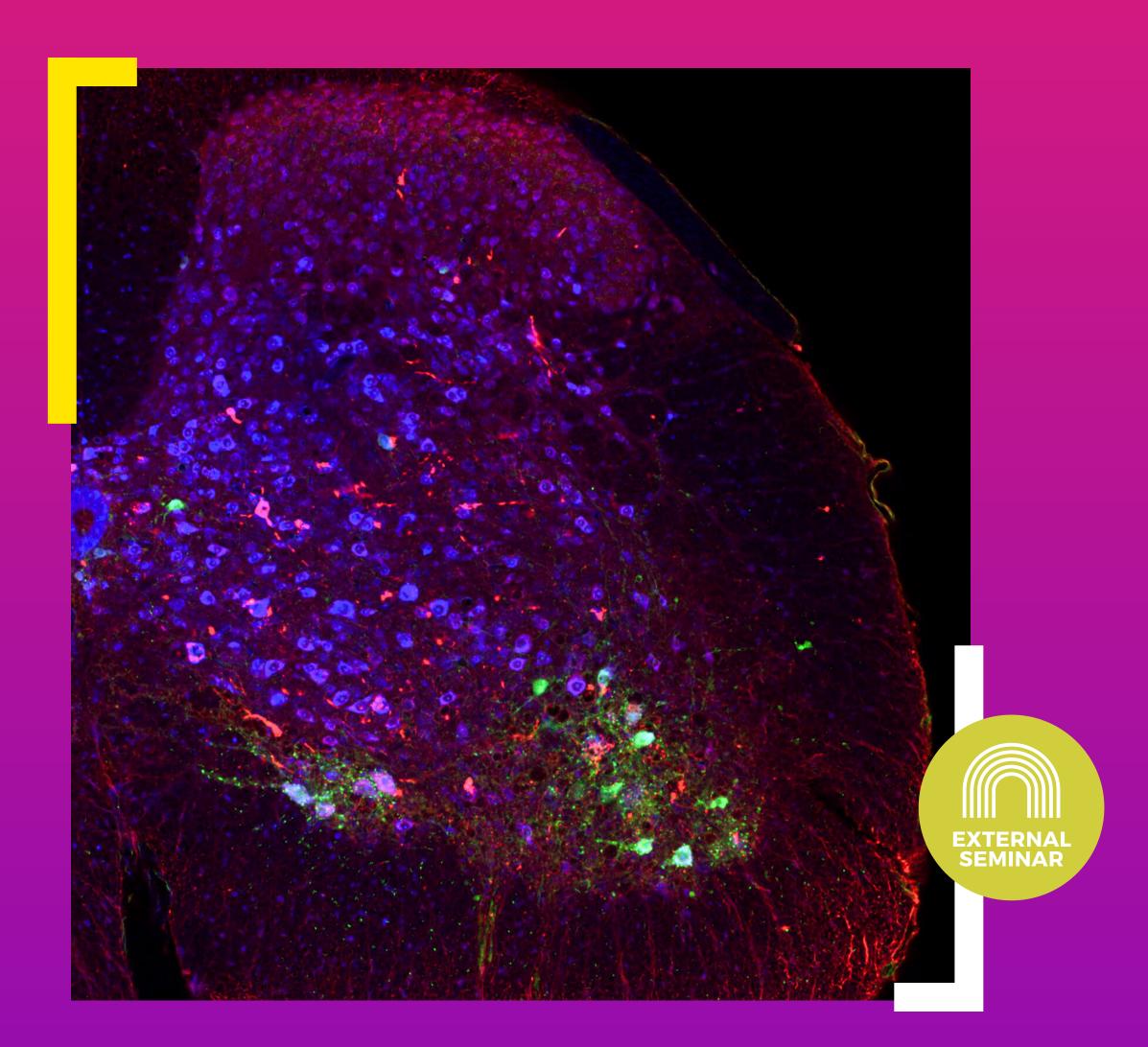


19 SEPTEMBER

11.30 A.M. ROOM A106 - POVO 1

ENTER FOR BRAIN RESEARCH



- TBK1: A DOUBLE-EDGED SWORD IN
- AMYOTROPHIC LATERAL SCLEROSIS
 - DISEASE PROGRESSION

DNA sequence variants in the TBK1 (TANK-Binding Kinase 1) gene associate with the neurodegenerative diseases Amyotrophic Lateral Sclerosis (ALS) and FrontoTemporal Dementia (FTD). We have generated and characterized mice bearing human ALS/FTD-associated TBK1 missense loss-of-function mutations. We observed that TBK1 mutations affect distinct intracellular pathways (autophagy and the Type-Interferon response) in specific cell types (motor neurons and glia), and at different stages of disease progression. Our work further highlighted a key role of the TBK1-dependent Type-I Interferon response in driving ALS disease severity in mice and humans, and highlighted its potential as a therapeutic target to slow neurodegenerative disease progression.