Names (Presenter in bold font)	**Wei Luan**, Amanda Wright, Heledd Brown-Wright, Rebecca San Gil, Lidia Madrid San Martin, Adam Walker
Affiliations	**Queensland Brain Institute, the University of Queensland**
Title	Early activation of cell stress and death pathways due to TDP-43 pathology in rNLS mouse model for ALS
Abstract (max 300w)	**The rNLS8 human TDP-43 (hTDP-43) transgenic mouse model shows cytoplasmic accumulation of TDP-43 and hyperphosphorylated TDP-43 that are pathological hallmarks for ALS/FTD, recapitulating neuronal loss, progressive motor dysfunction and eventual death due to the disease. However, it remains unsolved how TDP-43 pathology impacts the biological pathways key to homeostasis in neurons, consequently leading to neuronal death. In this study, we employed a customised RT2 qPCR array to examine the expression profiles of pre-selected 71 genes that play essential roles in several important biological pathways contributing to ALS/FTD. The array data show significant upregulation of several genes belonging to several pathways of the integrated stress response, oxidative stress, DNA damage response and apoptosis in the cortex of rNLS mice at early disease onset compared to non-transgenic littermates. We also identified two downregulated genes involved in cellular energy and intracellular pH maintenance, two cellular metabolic pathways critical for neuronal survival. Furthermore, these altered genes identified in the RT2 array were validated using real-time qPCR. In summary, our finding revealed early dysregulation of genes in several vital pathways in response to TDP-43 pathology in early disease, offering new therapeutic targets for the treatment of ALS/FTD.**

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