Dantrolene protects neurons from Huntington's disease

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Huntington's disease (HD) is characterized by ongoing destruction of specific neurons within the brain. It affects a person's ability to walk, talk, and think - leading to involuntary movement and loss of muscle co-ordination. New research published in BioMed Central's open access journal *Molecular Neurodegeneration* shows that the RyanR inhibitor Dantrolene is able to reduce the severity of walking and balance problems in a mouse model of HD.

Progressive damage to medium spiny neurons (MSN) in the brain of a person with HD is responsible for many of the symptoms and is caused by an inherited recessive mutation in the gene 'Huntingtin'. The mutated version of this protein leads to abnormal release of calcium from stores within the neurons which in turn disrupts the connections between neurons firing and muscle contractions, and eventually kills the neurons.

Researchers from the University of Texas Southwestern Medical Center tested Dantrolene, a muscle relaxant which works by stabilizing calcium signaling, and showed that this drug could prevent calcium-dependent toxicity in laboratory grown neurons. The team led by Dr Ilya Bezprozvanny also found that Dantrolene could prevent destruction of co-ordination, measured by beam walking and footprint patterns, in mice with Huntington's-like disease.

Dr Bezprozvanny explained, "One of the features of HD mice is the progressive loss of their NeuN-positive neurons. Dantrolene was not only able to protect muscle co-ordination in mice with HD but also prevented

destruction of NeuN positive neurons. Our results suggest that RyanR inhibitors, such as Dantrolene, should be considered as future treatments to slow down the effects of diseases like Huntington's."

More information: Dantrolene is neuroprotective in Huntingtons disease transgenic mouse model Xi Chen, Jun Wu, Svetlana Lvovskaya, Emily Herndon, Charlene Supnet and Ilya Bezprozvanny. *Molecular Neurodegeneration* (in press)

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