Mitochondrial dysfunction in Huntington’s disease: what went wrong, when and where?

Huntington’s disease (HD) is an inherited, neurodegenerative disorder affecting adults with loss of motor control, psychiatric symptoms and cognitive decline. Despite knowledge of the disease-causing mutation, and well-described pathology, the mechanisms behind the pathogenesis are not yet fully elucidated. In both HD patients and some HD animal models a pronounced weight loss is seen in the late stages of the disease, which has led to the hypothesis that alterations in energy metabolism play a role in HD. Interestingly, the Medium Spiny Neurons in the striatum, who are selectively lost in HD, also display high vulnerability to treatment with mitochondrial toxins, indicating that loss of normal mitochondrial function in these neurons could be of major importance in HD pathogenesis. We and others have pursued this line of investigation; in the lecture I will review the literature and present our own published and unpublished results.

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Guests are welcome