Mutant protein spread in Huntington’s disease and its implications for other neurodegenerative disorders of the CNS.

https://neurodegenerationresearch.eu/survey/mutant-protein-spread-in-huntingtons-disease-and-its-implications-for-other-neurodegenerative-disorders-of-the-cns/

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5

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Research Abstract
Huntington’s disease (HD) is a rare inherited disorder in which the abnormal gene codes for a protein (mutant huntingtin, mHtt) that over time accumulates in the brain, killing nerve cells and causing disease. This disease typically occurs in mid life, progresses to death over 10-30 years and is incurable. To date the problem in HD has been thought to be due to nerve cells producing their own, genetically coded for, abnormal mHtt which then causes them to

dysfunction and die. In recent years, there has been evidence to suggest that other cells, such as those involved in inflammation, may also contribute to the loss of neuronal cells. However, such cells have only been thought of as having indirect effects and not central to the disease process. Recently though, we have found in HD patients – who were transplanted with fetal tissue designed to replace cells lost to the disease process – that the abnormal mHtt could be seen in the transplant. This mHtt could only have got into the transplant from the patient as the grafted tissue was from unaffected donors. This unique observation forms the basis of this application as we now investigate the theory that mHtt can be transferred between cells either via host nerve cell connections and/or from circulating immune cells that can deliver the mHtt into the brain via inflamed, leaky blood vessels. This will be investigated using a range of techniques including studies with animals genetically engineered to carry the HD gene, cell cultures, state-of-the-art brain imaging techniques that allow to visualize cells within living tissue in real time, and blood and brain samples from HD patients. If true this new theory would have wide ranging implications not just for HD, but other similar diseases, and bring with it a totally new therapeutic approach to this currently incurable condition.

Further information available at:

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