

Huntington's disease problem start early

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The damaging effects of the mutated protein involved in Huntington's disease take place earlier in cell life than previously believed, said researchers from Baylor College of Medicine in Houston in a report that appears in the current edition of the journal *Neuron*.

“This research provides evidence of toxicity by huntingtin (the protein involved in the disease) early during the disease process,” said Dr. Juan Botas, associate professor of molecular and human genetics at BCM. Previously, researchers thought that the protein, which is extremely large, begins its negative effects after it is cut and imported into the cell's nucleus. However, Botas and his colleagues showed that the toxic effects are felt even before the protein is cleaved.

“Early in the disease, the full-length protein already causes neurotransmission problems at the level of the synapse,” he said. The synapse allows communication between two cells using chemicals called neurotransmitters. “We investigated the nature of those neurotransmitter defects, and at the same time, identified the genes that could ameliorate those defects.”

Huntington's disease is one of nine diseases associated with a repeat of the three nucleotides [C (cytosine) A (adenine) G (guanine)]. The combination of the three make an amino acid called glutamine. When the CAG repeats an inordinate amount of time, it results in a disease. Collectively, the disorders are called polyglutamine diseases.

Among those genes that can compensate for the misfire at the synapse

are those that govern calcium channels (tiny pores in the cell's membrane that allow the in- and out-flow of calcium), said Botas. These genes and others involved in synaptic transmission could serve as targets for the development of potential drug therapies.

Source: Baylor College of Medicine

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