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Researchers show the transmission of the genetic disorder HD in normal animals

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Mice transplanted with cells grown from a patient suffering from Huntington's disease (HD) develop the clinical features and brain pathology of that patient, suggests a study published in the latest issue of *Acta Neuropathologica* by CHA University in Korea, in collaboration with researchers at Université Laval in Québec City, Canada.

"Our findings shed a completely new light onto our current understanding of how HD begins and develops. We believe that they will also lead to the development of a whole new range of therapies for neurodegenerative diseases of the central nervous system", explains corresponding author of the study Jihwan Song, professor and director of Neural Regeneration and Therapy Group at the CHA Stem Cell Institute of CHA University.

The researchers have now provided further evidence for this new theory by showing that the abnormal protein coded for this genetic disorder can be transmitted to normal animals by the injection of diseased cells into their brain. "This is the first demonstration that cells carrying a genetic disease are capable of spreading into the normal mammalian brain and lead to the manifestation of behavioral abnormalities associated with the disease", says Francesca Cicchetti, professor at the Université Laval Faculty of Medecine and researcher at Centre de recherche du CHU de Québec-Université Laval.

HD is an inherited chronic degenerative disorder of the brain characterized by major thinking and motor problems as well as

psychiatric disturbances. There is no cure for HD and current treatments are of very limited efficacy. It is caused by a single gene abnormality which leads to the production of a mutant form of a protein called huntingtin (mHtt). The production of this protein in a nerve cell eventually kills it but it has long been thought that this protein cannot spread out of the cell and infect and kill neighbouring ones.

However, in recent post mortem analyses of HD patients who received transplants of non-HD tissue in an attempt to repair their brain, the researchers showed that the mHtt can be found in the graft itself. This suggests that the patient with HD transmitted the mHtt from their brain into the transplant.

Provided by Laval University