Rosa HD

Tech ID: 21529 / UC Case 2009-522-0

BACKGROUND

Huntington's disease (HD) is an adult-onset, autosomal-dominant neurodegenerative disease that is clinically characterized by a triad of movement disorders (i.e., chorea and bradykinesia), psychiatric symptoms, and cognitive deficits. In afflicted patients, symptoms usually progress relentlessly until death in 15-20 years after disease onset. HD is one of nine neurodegenerative disorders caused by a CAG repeat expansion encoding a polyglutamine (polyQ) repeat in otherwise unrelated proteins. In HD, the mutated *Huntingtin* (*mhtt*) protein is ubiquitously expressed in both neuronal and nonneuronal tissues. The polyQ repeat, located in the N terminus of huntingtin (htt), is normally less than 36, but is expanded to more than 37 in HD patients. In all polyQ disorders, there is an inverse relationship between the length of polyQ and the age of disease onset. Currently, there is no effective treatment or cure for HD or any other polyQ disorder.

INNOVATION

Researchers at UCLA have developed a Cre/LoxP conditional mouse model of HD (termed RosaHD mice) in which expression of a toxic mutant huntingtin Exon 1 (mhtt-exon1) fragment, driven by the endogenous ubiquitously-expressing Rosa26 promoter, can be switched on by Cre recombinase.

APPLICATIONS

- Useful in studying Huntington's disease (HD) or other polyQ disorders.

RELATED MATERIALS

- Pathological cell-cell interactions elicited by a neuropathogenic form of mutant Huntingtin contribute to cortical pathogenesis in HD mice
- Pathological cell-cell interactions are necessary for striatal pathogenesis in a conditional mouse model of Huntington's disease
- Information on Mouse Strain from The Jackson Laboratory

OTHER INFORMATION

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ADDITIONAL TECHNOLOGIES BY THESE INVENTORS

- Novel Mouse Model for Huntingtons Disease
- Transgenic Mouse Model of Parkinson's Disease with Age-Dependent Hypokinetic Motor Deficits, Dopaminergic Neuron Loss, and Alpha Synuclein Accumulation

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OTHER INFORMATION

CATEGORIZED AS

- **Medical**
  - Disease: Central Nervous System
- **Research Tools**
  - Animal Models

RELATED CASES

2009-522-0

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