BULLETIN

CANADIAN GENETIC DISEASES NETWORK

# SCIENTISTS DEVELOP FIRST MOUSE MODEL TO REPRODUCE CARDINAL FEATURES OF HUNTINGTON DISEASE

**Vancouver - May 27, 1999:** A team of researchers at the Centre for Molecular Medicine and Therapeutics, led by Dr. Michael Hayden, Scientific Director of the Canadian Genetic Diseases Network, has spearheaded a breakthrough in the study of the neurological disease, Huntington disease (HD). The scientists, in collaboration with colleagues at the University of British Columbia, University of Toronto, and in the United States have produced yeast artifical chromosome (YAC) transgenic mice expressing the cardinal features of HD.

Huntington disease is associated with a gradual deterioration of brain cells which leads to onset of personality change, involuntary movements, and dementia typically between the ages of 35 and 45, and eventual death.

In the study, the team produced mice expressing normal and mutant huntingtin protein in a developmental and tissue-specific manner identical to that observed in Huntington disease. The mutant mice show early electrophysiological abnormalities indicating cytoplasmic dysfunction prior to indications of neurodegeneration, then show selective degeneration of those specific neuronal cells that die in patients with HD. The results of the study are published today in the scientific journal *Neuron.*

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Dr. Hayden commented that the new YAC mouse allows scientists to dissect the natural history of HD. In producing animals with the behavioral, electrophysiological and

pathological characteristics of different stages of development of the disease, his team now has insights into the sequence of cellular and molecular events underlying the pathogenesis of HD. The discovery, which was supported by CGDN, the Medical Research Council of Canada, the Huntington Disease Society of America, and the National Institute of Health, will allow scientists to assess therapeutic strategies to delay or prevent the disease.

The Centre for Molecular Medicine and Therapeutics is a core research centre of the Children's and Women's Health Centre of B.C. and the Canadian Genetic Diseases Network. It is an innovative and unique partnership among Merck Frosst Canada Inc., the government of B.C., the University of British Columbia, and the Health Centre dedicated to the conduct of advanced research using novel approaches in molecular and cell biology and genetics, to obtain fundamental advances in knowledge.

The Canadian Genetic Diseases Network is a not-for-profit nation-wide centre of research excellence under the federal Networks of Centres of Excellence Program. CGDN creates strategic partnerships to commercialize discoveries for the benefit of health care in Canada and worldwide. Fifty scientists and their teams, based in 18 universities, hospitals, and research centres across Canada, focus on the molecular and cellular causes of inherited disease. In 1997, CGDN was awarded a further seven years' core funding by the Networks of Centres of Excellence Program via the Medical Research Council of Canada, to conduct an expanded research program "From Genes to Therapies."

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