NEURODEGENERATIVE DISEASES

Ref. 30743

HUNTINGTON´S DISEASE (HD)

Green Fluorescent mHTT-92Q (exon 1) inducible CELL LINE

Huntington's disease (HD) is a hereditary progressive autosomal dominant neurodegenerative disease characterized by choreatic movements and neuropsychiatric disorders leading to cognitive decline.

HD is caused as a result of a mutation that multiplies the CAG codons encoding the amino acid glutamine in the exon 1 on the short arm of chromosome 4p16.3 in the Huntingtine (HTT) gene.

The number of CAG triplets can vary over a wide range. The disease developes when the number of repetitions exceeds 35.

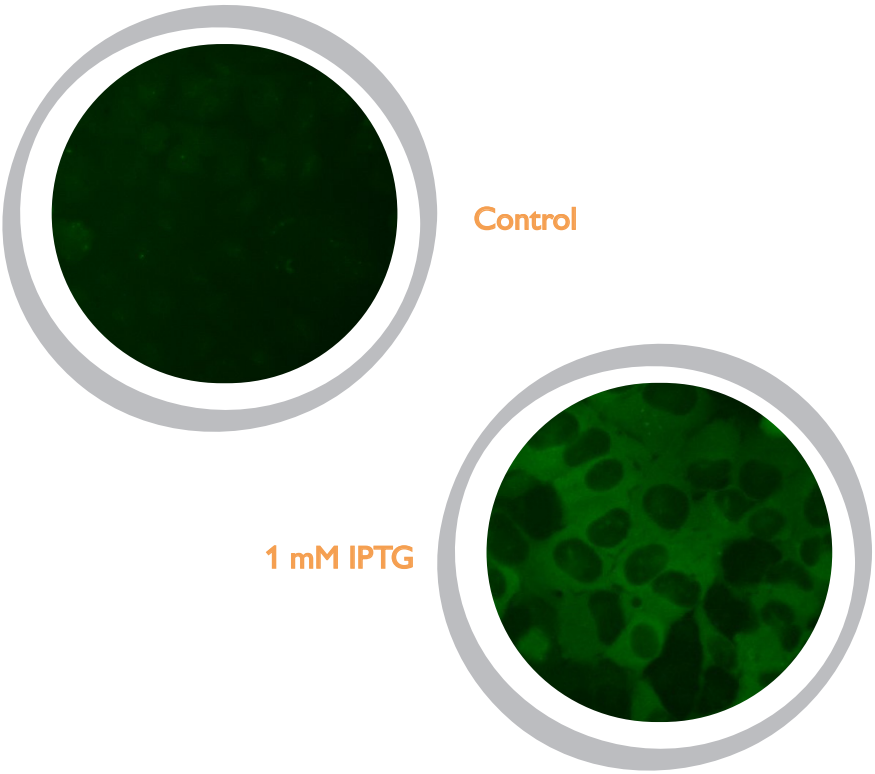
The longer the CAG repeats, the earlier the onset of disease. In cases of juvenile onset Huntingtine´s disease (JHD) the CAG triplets often exceeds 55.

Innoprot

Background

Mutant HTT (mHTT) fragments have been described to underlie the molecular pathogenesis of Huntington´s disease. The toxicity of N-terminal fragments has been studied in human post-mortem tissues and mouse models concluding that the exon 1 of mutant HTT protein is the most pathogenic HTT fragment. Indeed, the aggregates have been only detected by antibodies to the N-terminus of HTT.

Innoprot´s green fluorescent mHTT-92Q (exon 1) inducible U2OS cell line stably expresses a green fluorescent tagged mutant fragment of HTT exon 1 containing 92 CAG (glutamine coding) triplets under the Lac operon expression control.



Product Name: Green Fluorescent mHTT-92Q (exon 1) inducible cell line
Product reference: P30743
Prot. Official Full Name: mHTT-92Q (exon 1)
Host Cell: U2OS
Resistance: Puromycin and Hygromycin
Quantity: > 3 x 10^6 cells / vial
Storage: Liquid Nitrogen