Revision 4

#2773 Store at -20C

Huntingtin Antibody

Cell Signaling TECHNOLOGY®

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For Research Use Only. Not for Use in Diagnostic Procedures.

Applications:	Reactivity:	Sensitivity:	MW (kDa):	Source/Isotype:	UniProt ID:	Entrez-Gene Id:
W, IP, IF-F	H M R	Endogenous	350	Rabbit	#P42858	3064

Product Usage Information

Application	Dilution
Western Blotting	1:1000
Immunoprecipitation	1:50
Immunofluorescence (Frozen)	1:100

Storage

Supplied in 10 mM sodium HEPES (pH 7.5), 150 mM NaCl, 100 µg/ml BSA and 50% glycerol. Store at –20°C. Do not aliquot the antibody.

Specificity/Sensitivity

Huntingtin Antibody detects endogenous levels of total huntingtin protein.

Source / Purification

Polyclonal antibodies are produced by immunizing animals with a synthetic peptide corresponding to residues surrounding Pro1218 of human huntingtin protein. Antibodies are purified by protein A and peptide affinity chromatography.

Background

Huntington's Disease (HD) is a fatal neurodegenerative disorder characterized by psychiatric, cognitive, and motor dysfunction. Neuropathology of HD involves specific neuronal subpopulations: GABA-ergic neurons of the striatum and neurons within the cerebral cortex selectively degenerate (1,2). The genetic analysis of HD has been the flagship study of inherited neurological diseases from initial chromosomal localization to identification of the gene.Huntingtin is a large (340-350 kD) cytosolic protein that may be involved in a number of cellular functions such as transcription, gastrulation, neurogenesis, neurotransmission, axonal transport, neural positioning, and apoptosis (2,3). The HD gene from unaffected individuals contains between 6 and 34 CAG trinucleotide repeats, with expansion beyond this range causing the onset of disease symptoms. A strong inverse correlation exists between the age of onset in patients and the number of huntingtin gene CAG repeats encoding a stretch of polyglutamine peptides (1,2). The huntingtin protein undergoes numerous post-translational modifications including phosphorylation, ubiquitination, sumoylation, palmitoylation, and cleavage (2). Phosphorylation of Ser421 by Akt can partially counteract the toxicity that results from the expanded polyglutamine tract. Varying Akt expression in the brain correlates with regional differences in huntingtin protein phosphorylation; this pattern inversely correlates with the regions that are most affected by degeneration in diseased brain (2). A key step in the disease is the proteolytic cleavage of huntingtin protein into amino-terminal fragments that contain expanded glutamine repeats and translocate into the nucleus. Caspase mediated cleavage of huntingtin at Asp513 is associated with increased polyglutamine aggregate formation and toxicity. Phosphorylation of Ser434 by CDK5 protects against cleavage (2,3).

Background References

1. Gusella, J.F. and Macdonald, M.E. (2006) *Trends Biochem. Sci.* 31, 533-540.
2. Borrell-Pagès, M. et al. (2006) *Cell Mol. Life Sci.* 63, 2642-2660.
3. Luo, S. et al. (2005) *J. Cell Biol.* 169, 647-656.

Species Reactivity

Species reactivity is determined by testing in at least one approved application (e.g., western blot).

Western Blot Buffer

IMPORTANT: For western blots, incubate membrane with diluted primary antibody in 5% w/v BSA, 1X TBS, 0.1% Tween® 20 at 4°C with gentle shaking, overnight.

Applications Key

W: Western Blotting **IP:** Immunoprecipitation **IF-F:** Immunofluorescence (Frozen)

Cross-Reactivity Key

H: Human **M:** Mouse **R:** Rat

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Limited Uses

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