

Store at
-20°C

Huntingtin (D7F7) XP[®] Rabbit mAb

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#5656

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orders@cellsignal.comEntrez-Gene ID #3064
UniProt ID #P42858

rev. 09/25/18

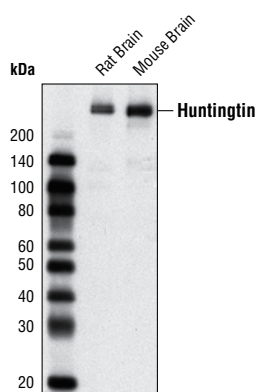
For Research Use Only. Not For Use In Diagnostic Procedures.

| Applications | Species Cross-Reactivity* | Molecular Wt. | Isotype |
|----------------------------------|---------------------------|---------------|------------|
| W, IP, IHC-P, IF-F Endogenous | H, M, R | 350 kDa | Rabbit IgG |

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).

Confocal immunofluorescent analysis of rat hippocampus using ► Huntingtin (D7F7) XP[®] Rabbit mAb (green). Red = propidium iodide, a fluorescent DNA dye.

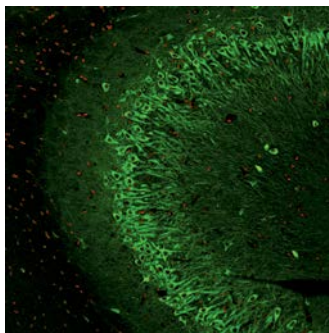


Western blot analysis of extracts from rat and mouse brain tissue using Huntingtin (D7F7) XP[®] Rabbit mAb.

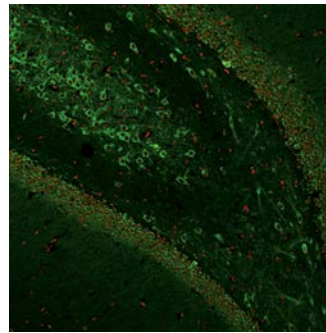
Specificity/Sensitivity: Huntingtin (D7F7) XP[®] Rabbit mAb detects endogenous levels of total huntingtin protein. Species cross-reactivity for IHC-P is in rodent only.

Source/Purification: Monoclonal antibody is produced by immunizing animals with a synthetic peptide corresponding to residues surrounding Pro1220 of human huntingtin protein.

CA3



Dentate Gyrus



Storage: Supplied in 10 mM sodium HEPES (pH 7.5), 150 mM NaCl, 100 µg/ml BSA, 50% glycerol and less than 0.02% sodium azide. Store at -20°C. Do not aliquot the antibody.

*Species cross-reactivity is determined by western blot.

**Anti-rabbit secondary antibodies must be used to detect this antibody.

Recommended Antibody Dilutions:

| | |
|--|--|
| Western blotting | 1:1000 |
| Immunoprecipitation | 1:50 |
| Immunohistochemistry (Paraffin) | 1:800 |
| Unmasking buffer: | Citrate |
| Antibody diluent: | SignalStain [®] Antibody Diluent #8112 |
| Detection reagent: | SignalStain [®] Boost (HRP, Rabbit) #8114 |
| Optimal IHC dilutions determined using SignalStain [®] Boost IHC Detection Reagent. | |
| Immunofluorescence (IF-F) | 1:100 |

For product specific protocols and a complete listing of recommended companion products please see the product web page at www.cellsignal.com.

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.

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

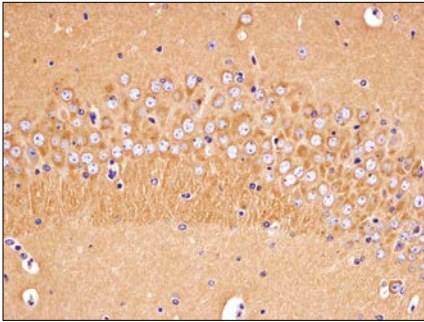
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Applications: W—Western IP—Immunoprecipitation IHC—Immunohistochemistry ChIP—Chromatin Immunoprecipitation IF—Immunofluorescence F—Flow cytometry E-P—ELISA-Peptide Species Cross-Reactivity: H—human M—mouse R—rat Hm—hamster Mk—monkey Mi—mink C—chicken Dm—D. melanogaster X—Xenopus Z—zebrafish B—bovine Dg—dog Pg—pig Sc—S. cerevisiae Ce—C. elegans Hr—Horse All—all species expected Species enclosed in parentheses are predicted to react based on 100% homology.

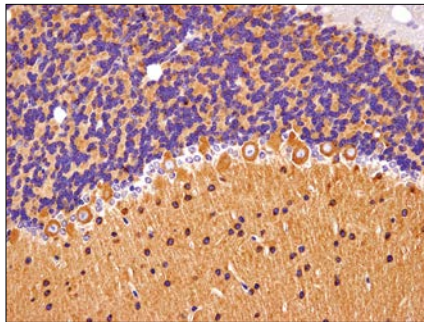
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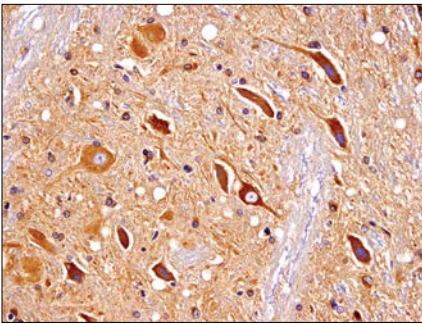
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Immunohistochemical analysis of paraffin-embedded mouse brain using Huntingtin (D7F7) XP® Rabbit mAb.



Immunohistochemical analysis of paraffin-embedded mouse cerebellum using Huntingtin (D7F7) XP® Rabbit mAb.



Immunohistochemical analysis of paraffin-embedded rat brain using Huntingtin (D7F7) XP® Rabbit mAb.