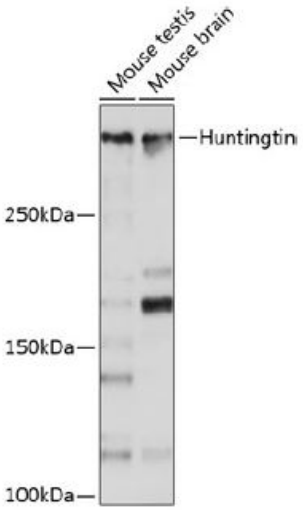
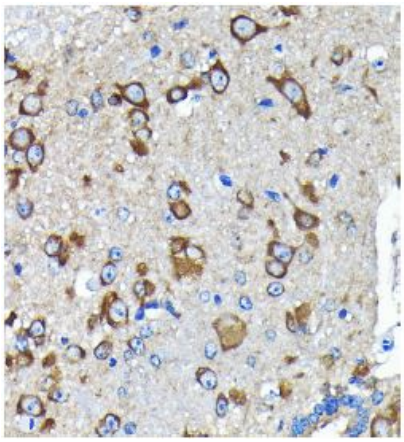
Product name:	HuntingtIn
Cat number:	MAB-94596
Conjugate:	Unconjugated
Size:	100 ug
Clone:	D7F7
Concentration:	1mg/ml
Host:	Rb
Isotype:	IgG
Reactivity:	Hu, Ms, Rt
Applications:	WB, IHC, IF Western Blot: 1:500-1:2000 Immunohistochemistry (Paraffin): 1:50-1:200 Immunofluorescence: 1:50-1:200
Molecular Weight:	350 kDa
Purification:	Monoclonal antibody is produced by immunizing animals with a synthetic peptide corresponding to residues surrounding Pro1220 of human huntingtin protein.
Form:	Liquid
Buffer:	PBS with 0.02% sodium azide, 0.05% BSA,50% glycerol, pH7.3.
Storage:	Store at -20℃. Avoid freeze / thaw cycles.

Background:

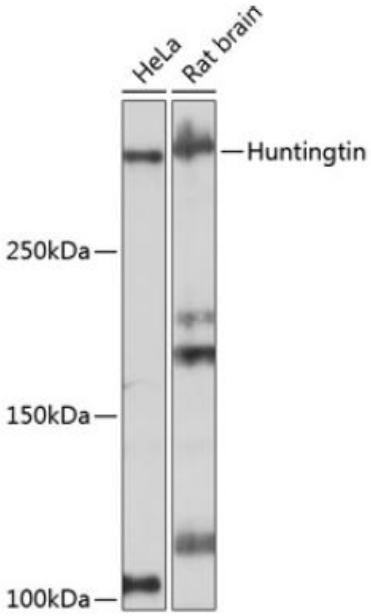
Huntington's Disease (HD) is a fatal neurodegenerative disorder characterized by psychiatric, cognitive, and motor dysfunction. Neuropathology of HD involves specific neuronal subpopulations: GABA-ergic neurons of the striatum and neurons within the cerebral cortex selectively degenerate (1,2). The genetic 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). Huntingtin (D7F7) XP® Rabbit mAb detects endogenous levels of total huntingtin protein. Species cross-reactivity for IHC-P is in rodent only..



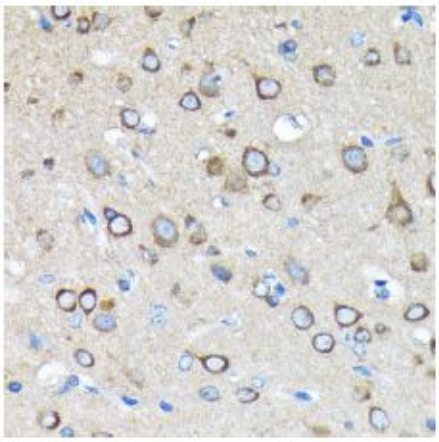
Western blot analysis of extracts of various cell lines, using Huntingtin antibody at 1:1000 dilution. Secondary antibody: HRP Goat Anti- Rabbit IgG (H+L) at 1:10000 dilution. Lysates/proteins: 25ug per lane. Blocking buffer: 3% nonfat dry milk in TBST. Detection: ECL West Pico Plus. Exposure time: 10s.



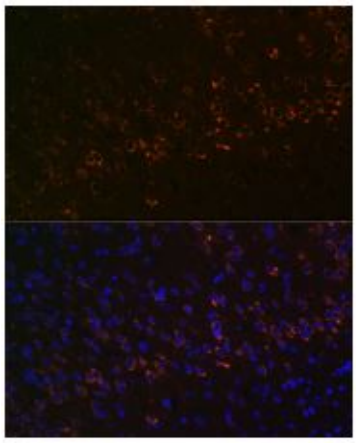
Immunohistochemistry of paraffinembedded mouse spinal cord using Huntingtin Rabbit mAb at dilution of 1:100 (40x lens).



tfistern blot analysis of extracts of various cell lines, using Huntingtin antibody at 1:1000 dilution. Secondary antibody: HRP Goat Anti-Rabbit IgG (H+L) at 1:10000 dilution. Lysates/proteins: 25ug per lane. Blocking buffer: 3% nonfat dry milk in TBST. Detection: ECL West Pico Plus. Exposure time: 3min.



Immunohistochemistry of paraffinembedded rat brain using Huntingtin Rabbit mAb at dilution of 1:100 (40x lens).



Immunofluorescence analysis of mouse brain using Huntingtin Rabbit mAb at dilution of 1:100 (40x lens). Blue: DAPI for nuclear staining.