SunLong Biotech Co.,LTD
Tel: 0086-571- 56623320 Fax:0086-571- 56623318
E-mail:sales@sunlongbiotech.com
www.sunlongbiotech.com

Rabbit Anti-Huntingtin antibody

SL11699R

Product Name:	Huntingtin
Chinese Name:	神经性舞蹈病蛋白抗体
Alias:	HD; HD protein; HD_HUMAN; HDH; HTT; Huntingtin; HUNTINGTON CHOREA; Huntington disease protein; Huntington's disease protein homolog; IT 15; IT15; OTTMUSP00000026909; ZHD; AI256365; C430023I11Rik.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Pig,Cow,Horse,Rabbit,Sheep,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	347kDa
Cellular localization:	The nucleuscytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Huntingtin:751-850/3142
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	Huntingtin is a protein that contains a polyglutamine region. When the number of glutamine repeats exceeds 35, the gene encodes a version of Huntingtin that leads to Huntington’s disease (HD). When the polyglutamine stretch is mutated, Huntingtin acts within the nucleus to induce neurodegeneration by a cell-specific apoptotic mechanism. Loss of Huntingtin activity is unlikely to be the cause of HD, and it has been proposed

	that the expanded glutamine repeat region may induce an abnormal interaction between the mutant protein and other cellular proteins. Huntingtin interacts with a variety of proteins including HAP1, glyceraldehyde phosphate dehydrogenase (GAPDH) and HIP1. **Function:** May play a role in microtubule-mediated transport or vesicle function. **Subunit:** Binds SH3GLB1 (By similarity). Interacts through its N-terminus with PRPF40A. Interacts with PQBP1, SETD2 and SYVN. Interacts with PFN1. **Subcellular Location:** Cytoplasm. Nucleus. The mutant Huntingtin protein colocalizes with AKAP8L in the nuclear matrix of Huntington's disease neurons. **Tissue Specificity:** Expressed in the brain cortex (at protein level). Widely expressed with the highest level of expression in the brain (nerve fibers, varicosities, and nerve endings). In the brain, the regions where it can be mainly found are the cerebellar cortex, the neocortex, the striatum, and the hippocampal formation. **Post-translational modifications:** Cleaved by apopain downstream of the polyglutamine stretch. The resulting N-terminal fragment is cytotoxic and provokes apoptosis. Forms with expanded polyglutamine expansion are specifically ubiquitinated by SYVN1, which promotes their proteasomal degradation. **DISEASE:** Defects in HTT are the cause of Huntington disease (HD) [MIM:143100]. HD is an autosomal dominant neurodegenerative disorder characterized by involuntary movements (chorea), general motor impairment, psychiatric disorders and dementia. Onset of the disease occurs usually in the third or fourth decade of life and symptoms progressively worsen leading to death in 10 to 20 years. Onset and clinical course depend on the degree of poly-Gln repeat expansion, longer expansions resulting in earlier onset and more severe clinical manifestations. HD affects 1 in 10,000 individuals of European origin. Neuropathology of Huntington disease displays a distinctive pattern with loss of neurons, especially in the caudate and putamen (striatum). **Similarity:** Belongs to the huntingtin family. Contains 10 HEAT repeats. **SWISS:** P42858

	Gene ID: 3064 **Database links:** Entrez Gene: 3064Human Entrez Gene: 15194Mouse Entrez Gene: 29424Rat Omim: 143100Human Omim: 613004Human SwissProt: P42858Human SwissProt: P42859Mouse SwissProt: P51111Rat Unigene: 518450Human Unigene: 209071Mouse Unigene: 482929Mouse Unigene: 11193Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.