

Product datasheet

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ARG22449 anti-Huntingtin antibody [HDC8A4]

Package: 50 μg Store at: -20°C

Summary

Product Description Mouse Monoclonal antibody [HDC8A4] recognizes Huntingtin

This antibody recognizes with an epitope corresponding to the HDC region (2703 - 2911 amino acids) of the huntingtin protein. Mouse anti Huntingtin antibody, clone HDC8A4 detects a 350KDa band on western blots but also detects smaller degradation products of huntingtin. Clone HDC8A4 recognizes both denatured and native huntingtin in human brain. The combined use of Mouse anti Huntingtin antibody, clone HDC8A4, HDB4E10 and HDA3E10 demonstrate that huntingtin is enriched in neuronal

cells in the brain.

Tested Reactivity Hu, Ms, Rb

Tested Application IHC-Fr, IP, WB

Host Mouse

Clonality Monoclonal
Clone HDC8A4
Isotype IgG1

Target Name Huntingtin
Species Human

Immunogen Recombinant protein corresponding to amino acids 2703 - 2911 of huntingtin.

Conjugation Un-conjugated

Alternate Names Huntingtin; Huntington disease protein; HD protein; IT15; HD

Application Instructions

Application table	Application	Dilution
	IHC-Fr	Assay-dependent
	IP	Assay-dependent
	WB	Assay-dependent
Application Note	IHC-Fr: Increased cytoplasmic staining, relative to nuclear, has been reported using formaldehyde as a fixative compared with acetone/methanol, see Wilkinson et al. * The dilutions indicate recommended starting dilutions and the optimal dilutions or concentrations should be determined by the scientist.	

Properties

Form Liquid

Purification Purification with Protein G.

Buffer PBS and 0.09% Sodium azide

Preservative 0.09% Sodium azide

Concentration 1 mg/ml

Storage instruction For continuous use, store undiluted antibody at 2-8°C for up to a week. For long-term storage, aliquot

and store at -20°C or below. Storage in frost free freezers is not recommended. Avoid repeated freeze/thaw cycles. Suggest spin the vial prior to opening. The antibody solution should be gently mixed

before use.

Note For laboratory research only, not for drug, diagnostic or other use.

Bioinformation

Gene Symbol HTT

Gene Full Name huntingtin

Background Huntingtin is a disease gene linked to Huntington's disease, a neurodegenerative disorder characterized

by loss of striatal neurons. This is thought to be caused by an expanded, unstable trinucleotide repeat in the huntingtin gene, which translates as a polyglutamine repeat in the protein product. A fairly broad range in the number of trinucleotide repeats has been identified in normal controls, and repeat numbers in excess of 40 have been described as pathological. The huntingtin locus is large, spanning 180 kb and consisting of 67 exons. The huntingtin gene is widely expressed and is required for normal development. It is expressed as 2 alternatively polyadenylated forms displaying different relative abundance in various fetal and adult tissues. The larger transcript is approximately 13.7 kb and is expressed predominantly in adult and fetal brain whereas the smaller transcript of approximately 10.3 kb is more widely expressed. The genetic defect leading to Huntington's disease may not necessarily eliminate transcription, but may confer a new property on the mRNA or alter the function of the protein. One candidate is the huntingtin-associated protein-1, highly expressed in brain, which has increased affinity for huntingtin protein with expanded polyglutamine repeats. This gene contains an upstream open reading frame in the 5' UTR that inhibits expression of the huntingtin gene product

through translational repression. [provided by RefSeq, Jul 2008]

Function May play a role in microtubule-mediated transport or vesicle function. [UniProt]

Calculated Mw 348 kDa

PTM Cleaved by apopain downstream of the polyglutamine stretch. The resulting N-terminal fragment is

cytotoxic and provokes apoptosis.

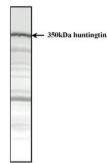
Forms with expanded polyglutamine expansion are specifically ubiquitinated by SYVN1, which

promotes their proteasomal degradation.

Phosphorylation at Ser-1179 and Ser-1199 by CDK5 in response to DNA damage in nuclei of neurons protects neurons against polyglutamine expansion as well as DNA damage mediated toxicity.

Images

human cerebral cortex



ARG22449 anti-Huntingtin antibody [HDC8A4] WB image

Western blot: Total protein extract of normal Human cerebral cortex separated as a strip on a 3-12.5% gradient SDS-PAGE gel and Western blotted. The blot was stained with ARG22449 anti-Huntingtin antibody [HDC8A4].