

Monoclonal Antibody to Huntingtin - Purified

Alternate names: HD, HTT, Huntington Disease Protein, IT15

Catalog No.: SM1661
Quantity: 0.1 mg
Concentration: 1.0 mg/ml

Background: Huntington's disease (HD) is a neurodegenerative disorder caused by an expanding

polyglutamine repeat in the huntingtin gene. HD is a mid-life onset autosomal dominant neurodegeneative disease that is characterized by psychiatric disorders, dementia, and involuntary movements (chorea), leading to death in 10-20 years. The HD gene product is widely expressed in human tissues, with the highest level of expression in the brain. The huntingtin gene product is expressed at similar levels in patients and controls, which suggests that the expansion of the polyglutamine repeat induces a toxic gain of function perhaps through interactions with other cellular proteins. Using yeast two-hybrid system, HAP1 (huntingtin associated protein 1) has been identified, that associates with huntingtin protein. The In vitro data suggest that the association between HAP1 and huntingtin is

enhanced by increasing length of glutamine repeat.

Uniprot ID: P42858

NCBI: <u>NP 002102.4</u>

GenelD: <u>3064</u>

Host / Isotype: Mouse / IgG1 Clone: HDB4E10

Immunogen: Recombinant protein corresponding to amino acids 1844 - 2131 of huntingtin

Format: State: Liquid purified IgG containing 0.09% Sodium Azide

Applications: Western blot: Clone HDB4E10 detects a 350KD band on western blots but also detects

smaller degradation products of huntingtin.

Immunoprecipitation.

Immunohistochemistry on frozen sections. Recommended positive control tissue: Brain.

Other applications not tested. Optimal dilutions are dependent on conditions and should

be determined by the user.

Specificity: This antibody reacts with an epitope corresponding to the HDB region (amino acids 1844 -

2131) of the huntingtin protein. The combined use of clone HDB4E10 (SM1661), HDC8A4 (SM1662) and HDA3E10 (SM1660) demonstrate that huntingtin is enriched in neuronal cells

in the brain.

Species: Rabbit, Human, Mouse.

Other species not tested.



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Storage:

Store the antibody undiluted at 2-8°C for one month or (in aliquots) at -20°C for longer.

Avoid repeated freezing and thawing. Shelf life: one year from despatch.

General References: 1. Wilkinson, F. L. et al. (1999) Localization of rabbit huntingtin using a new panel of

monoclonal antibodies. Molecular Brain Research. 69: 10-20.

Pictures:



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 probed with SM1661.