

Huntingtin Recombinant Rabbit mAb

Catalog: **BS48150** Host:

Rabbit

Reactivity: Human, Mouse, Rat

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 of trinucleotide repeats (9-35) 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 2016]

Product:

Store at -20 °C. Supplied in 50mM Tris-Glycine(pH 7.4), 0.15M NaCl, 40%Glycerol, 0.01% sodium azide and 0.05% BSA. Stable for 12 months from date of receipt.

Molecular Weight:

348 kDa

Swiss-Prot:

P42858

Purification&Purity:

Affinity Purification

Applications:

WB: 1:1000
IHC: 1:100
ICC/IF: 1:200
FC: 1:50

Storage&Stability:

Store at $4 \,^{\circ}{\rm C}$ short term. Aliquot and store at $-20 \,^{\circ}{\rm C}$ long term. Avoid freeze-thaw cycles.

Isotype:

IgG

DATA:

Note:

For research use only, not for use in diagnostic procedure.

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