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Monoclonal Antibody to Huntingtin

Anti human / mouse Huntingtin

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]

Product Number:	T-1441 (Lot 01PO1216)
Clone:	3HU-4E6
Host species, isotype:	Mouse IgG2b kappa
Quantity:	100µg
Format:	Affinity purified, lyophilized
	Reconstitute by adding 0.5ml distilled water. This stock solution contains 0.2mg/ml IgG, phosphate buffered saline pH 7.2 (PBS), 5mg/ml bovine serum albumin (BSA) as a stabilizer and 0.01% Kathon as a preservative.
Stability:	Original vial: 1 year at 4° - 8°C
	Stock solution or aliquots thereof: 1 year at -20°C. Avoid repeated thawing and freezing.
Applications:	Tested for immunohistochemistry (IHC). Not suitable for WB.
	Approximate working dilution for IHC: Frozen sections: not tested Paraffin sections: 2µg/ml (1:100); microwave pretreatment in citrate buffer is recommended for antigen retrieval.
	Optimal dilutions should be determined by the end user.

	Suggested positive control: human hypothalamus
	Please see **www.bma.ch** for protocols and general information.
Immunogen:	Recombinant Huntingtin fragment aa 1247 to 1646.
Antigen, epitope:	The antigen is Huntingtin mouse and human, the epitope lies in the region aa 1247 - 1646.
Antigen distribution:	Nucleus but also cytoplasmic expression in most tissues. Highest levels in neurons and paneth cells in gastrointestinal tract. Distinct granular expression pattern in several glandular epithelia.
Specificity:	**Human:** Huntingtin.
	Other: Cross-reactivity with mouse expected but not tested.

Selected references

Cellular localization of the Huntington's disease protein and discrimination of the normal and mutated form. Trottier Y, et al.: Nat Genet. 1995 May;10(1):104-10.

Polyglutamine expansion as a pathological epitope in Huntington's disease and four dominant cerebellar ataxias. Trottier Y, et al.: Nature. 1995 Nov 23;378(6555):403-6.

For *in vitro* research only. This product contains Kathon as a preservative.