

## Product Information

### Anti-Huntingtin (N-terminal)

produced in rabbit, affinity isolated antibody

Product Number **H7540**

### Product Description

Anti-Huntingtin (N-terminal) is produced in rabbit using as immunogen a synthetic peptide corresponding to a sequence at the N-terminal of human huntingtin (GeneID: 3064), conjugated to KLH. This sequence is identical in rat and mouse huntingtin. The antibody is affinity-purified using the immunizing peptide immobilized on agarose.

Anti-Huntingtin (N-terminal) specifically recognizes human huntingtin. The antibody may be used in various immunochemical techniques including immunoblotting (110 kDa). Detection of the huntingtin band by immunoblotting is specifically inhibited by the huntingtin immunizing peptide.

Huntington's disease (HD) is an autosomal dominant neurodegenerative disorder, characterized by progressive motor dysfunction, loss of cognitive function, and psychiatric disturbances, that inevitably leads to death.<sup>1,2</sup> Changes in the HD brain are widespread and include neuronal cell death and gliosis in the neocortex and striatum of HD patients. HD is caused by an aberrant expansion (>35) of glutamine repeats (polyQ) in the N-terminal region of huntingtin (htt), resulting in nuclear and cytoplasmic inclusion pathology. In addition, N-terminal proteolytic fragments of htt accumulate in the nucleus of affected neurons to form nuclear inclusions.<sup>3</sup> The precise cellular function of htt is not completely understood. Htt is essential for embryonic development and neurogenesis. Htt associates with various organelles, including the nucleus, endoplasmic reticulum and Golgi complex.<sup>4-6</sup> It is also found in neurites and synapses where it associates with vesicular structures and microtubules,<sup>6-8</sup> suggesting that it may play a role in intracellular vesicular trafficking, calcium homeostasis, neuronal survival, morphogenesis and transcriptional regulation.

### Reagent

Supplied as a solution in 0.01 M phosphate buffered saline, pH 7.4, containing 15 mM sodium azide as a preservative.

Antibody concentration: ~1.5 mg/mL

### Precautions and Disclaimer

For R&D use only. Not for drug, household, or other uses. Please consult the Safety Data Sheet for information regarding hazards and safe handling practices.

### Storage/Stability

For continuous use, store at 2–8 °C for up to one month. For extended storage, freeze in working aliquots at –20 °C. Repeated freezing and thawing, or storage in “frost-free” freezers, is not recommended. If slight turbidity occurs upon prolonged storage, clarify the solution by centrifugation before use. Working dilutions should be discarded if not used within 12 hours.

### Product Profile

**Immunoblotting:** a working antibody concentration of 0.5-1.0 µg/mL is recommended using a HEK-293T cell lysate expressing a N-terminal fragment of human huntingtin.

**Note:** In order to obtain best results in various techniques and preparations, it is recommended to determine optimal working dilutions by titration.

### References

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3. Schilling, G. et al., *J. Neuropathol. Exp. Pathol.*, **66**, 313-320 (2007).
4. DiFiglia, M. et al., *Neuron*, **14**, 1075-1081 (1995).
5. Veller, J. et al., *Exp. Neurol.*, **152**, 34-40 (1998).
6. Hoffner, G. et al., *J. Cell Sci.*, **115**, 941-948 (2002).
7. Gauthier, L.R. et al., *Cell*, **118**, 127-138 (2004).
8. Li, J.Y. et al., *Trends Mol. Med.*, **9**, 414-420 (2003).

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