

MATERIAL DATA SHEET**His₆-Ataxin-3 (MJD protein 1), human recombinant**
Cat. # E-341

Defects in the ATXN3 gene are the cause of Machado-Joseph disease (MJD), also known as spinocerebellar ataxia 3. MJD is an autosomal dominant neurodegenerative disorder caused by an expansion of a (CAG)_n repeat in the ATXN3 gene resulting in polymorphic protein forms in the C-terminal poly-glutamine region. Ataxin-3 protein belongs to a novel group of cysteine proteases similar to USP-type ubiquitin proteases and has deubiquitinating activity *in vitro*. The full-length protein contains an N-terminal Josephin domain, two ubiquitin interacting motifs (UIMs), and a variable C-terminus consisting of a polyglutamine stretch and tail. Ataxin-3 activity may have a role in MJD pathogenesis by influencing ubiquitin-dependent pathways that control protein folding and stability. This recombinant protein is N-terminally tagged. (Accession # NP_004984).

Product Information

Quantity:	50 µg
Stock:	X mg/ml (X µM) in 50 mM HEPES pH 8.0, 100 mM NaCl, 10% glycerol, 0.5 mM EDTA, 1mM DTT. Actual concentration varies with lot number.
MW:	42 kDa
Purity:	> 95% by SDS-PAGE

Use & Storage

Use:	Typical enzyme concentration for use <i>in vitro</i> ranges from 1-5 µM depending on conditions and substrate.
Storage:	Store at -80°C. Avoid multiple freeze/thaw cycles.

Literature

References:	Albrecht M., <i>et al</i> (2003) <u>Eur.J.Biochem.</u> 271 :3155-3170 Barrington B., <i>et al</i> (2003) <u>Hum.Mol.Genet.</u> 12 :3195-3205 Berke S. J., <i>et al</i> (2006) <u>J.Biol.Chem.</u> 280 :32026-32034 Chai Y., <i>et al</i> (2004) <u>J.Biol.Chem.</u> 279 :3605-3611 Donaldson K.M., <i>et al</i> (2003) <u>Proc.Natl.Acad.Sci.</u> 100 :8892-8897 Doss-Pepe E.W., <i>et al</i> (2003) <u>Mol.Cell.Biol.</u> 23 :6469-6483 Goto J., <i>et al</i> (1997) <u>Neurosci.Res.</u> 28 :373-377 Mao Y., <i>et al</i> (2005) <u>Proc.Natl.Acad.Sci.</u> 102 :12700-12705 Nijman S.M.B., <i>et al</i> (2005) <u>Cell.</u> 123 :773-786 Wang G., <i>et al</i> (2000) <u>Hum.Mol.Genet.</u> 9 :1795-1803
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