

Active Recombinant Human ATXN3, GST-tagged

Cat. No. ATXN3-173H Lot. No. (See product label)

SPECIFICATION

Product Overview	Recombinant human ATXN3 (amino acid residues 1-370), fused with N-terminal GST, was expressed in E.coli.
Species	Human
Source	E.coli
ProteinLength	1-370 a.a.
Description	<p>There are two main classes of DUB; cysteine proteases and metalloproteases. Ataxin-3 is a cysteine protease and is a member of the Machado-Joseph Domain (MJD) enzyme family. Cloning of the human gene was first described by Kawaguchi et al. (1994). Machado–Joseph disease (MJD), the most common form of spinocerebellar ataxia worldwide, is a progressive and ultimately fatal neurodegenerative disorder caused by polyQ expansion in ataxin-3, a conserved and ubiquitous protein known to bind polyubiquitin chains and to function as a deubiquitylating enzyme. Ataxin-3 has been linked to protein homeostasis maintenance, transcription, cytoskeleton regulation and myogenesis (Matos et al., 2011).</p>
Form	50 mM HEPES pH 7.5, 150 mM sodium chloride, 2 mM dithiothreitol, 10% glycerol
Bio-activity	Deubiquitylase Enzyme Assay: The activity of GST-Ataxin-3 was validated by determining the increase in fluorescence measured as a result of the enzyme catalysed cleavage of the fluorogenic substrate Ubiquitin-Rhodamine110-Glycine

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generating Ubiquitin and Rhodamine110-Glycine. Incubation of the substrate in the presence or absence of GST-Ataxin-3 was compared confirming the deubiquitylating activity of GST-Ataxin-3.

Molecular Mass	~69 kDa
Purity	>98% by SDS-PAGE
Storage	12 months at -70°C. Avoid multiple freeze/thaw cycles.
Concentration	0.5 mg/ml

GENE INFORMATION

Gene Name [ATXN3 ataxin 3 \[Homo sapiens \]](#)

Official Symbol ATXN3

Synonyms ATXN3; ataxin 3; Machado Joseph disease (spinocerebellar ataxia 3, olivopontocerebellar ataxia 3, autosomal dominant, ataxin 3) , MJD, SCA3; ataxin-3; ATX3; JOS; josephin; ataxin 3 variant h; ataxin 3 variant m; ataxin 3 variant ref; olivopontocerebellar ataxia 3; Machado-Joseph disease protein 1; spinocerebellar ataxia type 3 protein; Machado-Joseph disease (spinocerebellar ataxia 3, olivopontocerebellar ataxia 3, autosomal dominant, ataxin 3); AT3; MJD; MJD1; SCA3;

Gene ID [4287](#)

mRNA Refseq [NM_001127696](#)

Protein Refseq [NP_001121168](#)

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MIM	607047
UniProt ID	P54252
Chromosome Location	14q21
Pathway	Protein processing in endoplasmic reticulum, organism-specific biosystem; Protein processing in endoplasmic reticulum, conserved biosystem;
Function	cysteine-type peptidase activity; omega peptidase activity; peptidase activity; protein binding;

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