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AFG3L2 Antibody (N-term) Blocking Peptide

Synthetic peptide
Catalog # BP13219a

Specification

AFG3L2 Antibody (N-term) Blocking Peptide - Product Information

Primary Accession Q9Y4W6

AFG3L2 Antibody (N-term) Blocking Peptide - Additional Information

Gene ID 10939

Other Names
AFG3-like protein 2, 3424-, Paraplegin-like protein, AFG3L2

Target/Specificity
The synthetic peptide sequence used to generate the antibody AP13219a was selected from the N-term region of AFG3L2. A 10 to 100 fold molar excess to antibody is recommended. Precise conditions should be optimized for a particular assay.

Format
Peptides are lyophilized in a solid powder format. Peptides can be reconstituted in solution using the appropriate buffer as needed.

Storage
Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C.

Precautions
This product is for research use only. Not for use in diagnostic or therapeutic procedures.

AFG3L2 Antibody (N-term) Blocking Peptide - Protein Information

Name AFG3L2 {ECO:0000303|PubMed:10395799, ECO:0000312|HGNC:HGNC:315}

Function
Catalytic component of the m-AAA protease, a protease that plays a key role in proteostasis of inner mitochondrial membrane proteins, and which is essential for axonal and neuron development (PubMed:19748354, PubMed:28396416, PubMed:29932645, PubMed:30683687, PubMed:31327635, PubMed:37917749, PubMed:38157846). AFG3L2 possesses both ATPase and protease activities: the ATPase activity is required to unfold substrates, threading them into the internal proteolytic cavity for hydrolysis into small peptide fragments (PubMed:19748354, PubMed:<a

href="http://www.uniprot.org/citations/31327635" target="_blank">31327635). The m-AAA protease carries out quality control in the inner membrane of the mitochondria by mediating degradation of mistranslated or misfolded polypeptides (PubMed:26504172, PubMed:30683687, PubMed:34718584). The m-AAA protease complex also promotes the processing and maturation of mitochondrial proteins, such as MRPL32/bL32m, PINK1 and SP7 (PubMed:22354088, PubMed:29932645, PubMed:30252181). Mediates protein maturation of the mitochondrial ribosomal subunit MRPL32/bL32m by catalyzing the cleavage of the presequence of MRPL32/bL32m prior to assembly into the mitochondrial ribosome (PubMed:29932645). Required for SPG7 maturation into its active mature form after SPG7 cleavage by mitochondrial-processing peptidase (MPP) (PubMed:30252181). Required for the maturation of PINK1 into its 52kDa mature form after its cleavage by mitochondrial- processing peptidase (MPP) (PubMed:22354088). Acts as a regulator of calcium in neurons by mediating degradation of SMDT1/EMRE before its assembly with the uniporter complex, limiting the availability of SMDT1/EMRE for MCU assembly and promoting efficient assembly of gatekeeper subunits with MCU (PubMed:27642048, PubMed:28396416). Promotes the proteolytic degradation of GHITM upon hyperpolarization of mitochondria: progressive GHITM degradation leads to respiratory complex I degradation and broad reshaping of the mitochondrial proteome by AFG3L2 (PubMed:35912435). Also acts as a regulator of mitochondrial glutathione homeostasis by mediating cleavage and degradation of SLC25A39 (PubMed:37917749, PubMed:38157846). SLC25A39 cleavage is prevented when SLC25A39 binds iron-sulfur (PubMed:37917749, PubMed:38157846). Involved in the regulation of OMA1-dependent processing of OPA1 (PubMed:17615298, PubMed:29545505, PubMed:30252181, PubMed:30683687, PubMed:32600459). May act by mediating processing of OMA1 precursor, participating in OMA1 maturation (PubMed:29545505).

Cellular Location
Mitochondrion inner membrane; Multi-pass membrane protein

Tissue Location
Ubiquitous. Highly expressed in the cerebellar Purkinje cells.

AFG3L2 Antibody (N-term) Blocking Peptide - Protocols

Provided below are standard protocols that you may find useful for product applications.

- Blocking Peptides

AFG3L2 Antibody (N-term) Blocking Peptide - Images

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AFG3L2 Antibody (N-term) Blocking Peptide - Background

This gene encodes a protein localized in mitochondria andclosely related to paraplegin. The paraplegin gene is responsiblefor an autosomal recessive form of hereditary spastic paraplegia.This gene is a candidate gene for other hereditary spasticparaplegias or neurodegenerative disorders.

AFG3L2 Antibody (N-term) Blocking Peptide - References

Edener, U., et al. Eur. J. Hum. Genet. 18(8):965-968(2010)Di Bella, D., et al. Nat. Genet. 42(4):313-321(2010)Augustin, S., et al. Mol. Cell 35(5):574-585(2009)Mariotti, C., et al. Cerebellum 7(2):184-188(2008)Cagnoli, C., et al. Brain 129 (PT 1), 235-242 (2006) :