

AFG3L2 Antibody (N-term) Blocking Peptide
Synthetic peptide
Catalog # BP13219a**Specification**

AFG3L2 Antibody (N-term) Blocking Peptide - Product InformationPrimary 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 ([HGNC:315](#))**Function**

ATP-dependent protease which is essential for axonal and neuron development. In neurons, mediates 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). Required for paraplegin (SPG7) maturation (PubMed:30252181). After its cleavage by mitochondrial-processing peptidase (MPP), it converts paraplegin into a proteolytically active mature form (By similarity). Required for the maturation of PINK1 into its 52kDa mature form after its cleavage by mitochondrial-processing peptidase (MPP) (PubMed:22354088, PubMed:30252181). Involved in the regulation of OMA1-dependent processing of OPA1 (PubMed:32600459, PubMed:<a

[30252181](http://www.uniprot.org/citations/30252181)). Contributes to the proteolytic degradation of GHITM upon hyperpolarization of mitochondria (PubMed: [35912435](http://www.uniprot.org/citations/35912435)). Progressive GHITM degradation upon persistent hyperpolarization leads to respiratory complex I degradation and broad reshaping of the mitochondrial proteome by AFG3L2 (PubMed: [35912435](http://www.uniprot.org/citations/35912435)).

Cellular Location

Mitochondrion. Mitochondrion inner membrane {ECO:0000250|UniProtKB:Q8JZQ2}; 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**AFG3L2 Antibody (N-term) Blocking Peptide - Background**

This gene encodes a protein localized in mitochondria and closely related to paraplegin. The paraplegin gene is responsible for an autosomal recessive form of hereditary spastic paraplegia. This gene is a candidate gene for other hereditary spastic paraplegias 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) :