

AFG3L2 Antibody (N-term)
Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP13219a**Specification**

AFG3L2 Antibody (N-term) - Product Information

Application	IHC-P, WB,E
Primary Accession	O9Y4W6
Other Accession	O8JZO2 , O2KJ17 , NP_006787.2
Reactivity	Human
Predicted	Bovine, Mouse
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Calculated MW	88584
Antigen Region	52-80

AFG3L2 Antibody (N-term) - Additional Information**Gene ID** 10939**Other Names**

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

Target/Specificity

This AFG3L2 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 52-80 amino acids from the N-terminal region of human AFG3L2.

Dilution

IHC-P~~1:10~50

WB~~1:1000

E~~Use at an assay dependent concentration.

Format

Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.

Storage

Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions

AFG3L2 Antibody (N-term) is for research use only and not for use in diagnostic or therapeutic procedures.

AFG3L2 Antibody (N-term) - 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:[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

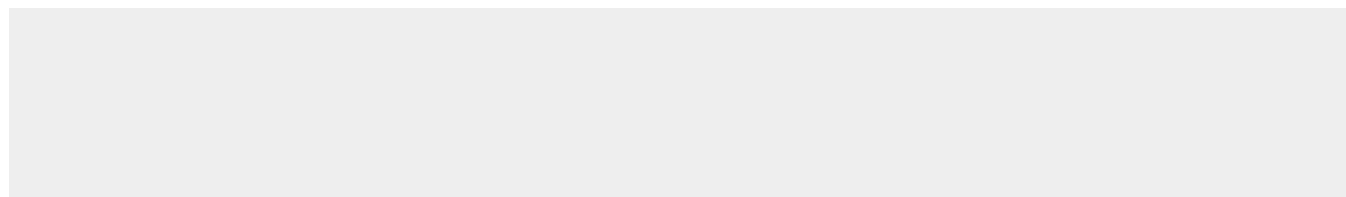
Ubiquitous. Highly expressed in the cerebellar Purkinje cells.

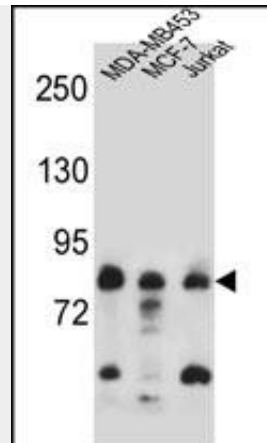
AFG3L2 Antibody (N-term) - Protocols

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

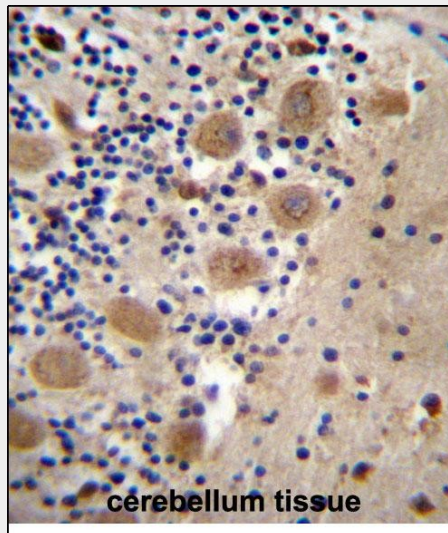
- [Western Blot](#)
- [Blocking Peptides](#)
- [Dot Blot](#)
- [Immunohistochemistry](#)
- [Immunofluorescence](#)
- [Immunoprecipitation](#)
- [Flow Cytometry](#)
- [Cell Culture](#)

AFG3L2 Antibody (N-term) - Images





AFG3L2 Antibody (N-term) (Cat. #AP13219a) western blot analysis in MDA-MB453, MCF-7, Jurkat cell line lysates (35ug/lane). This demonstrates the AFG3L2 antibody detected the AFG3L2 protein (arrow).



AFG3L2 Antibody (N-term) (Cat. #AP13219a) immunohistochemistry analysis in formalin fixed and paraffin embedded human cerebellum tissue followed by peroxidase conjugation of the secondary antibody and DAB staining. This data demonstrates the use of AFG3L2 Antibody (N-term) for immunohistochemistry. Clinical relevance has not been evaluated.

AFG3L2 Antibody (N-term) - 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) - 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) :

AFG3L2 Antibody (N-term) - Citations

- [Systematic analysis of a mitochondrial disease-causing ND6 mutation in mitochondrial deficiency](#)