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# AFG3L2 Antibody (N-term)

**Affinity Purified Rabbit Polyclonal Antibody (Pab)**
**Catalog # AP13219a**

## Specification

### AFG3L2 Antibody (N-term) - Product Information

| | |
|---|---|
| Application | **WB, IHC-P,E** |
| Primary Accession | Q9Y4W6 |
| Other Accession | Q8JZQ2, Q2KJI7, 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**
WB~~1:1000
IHC-P~~1:10~50

**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 (HGNC:315)

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**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:30252181). Contributes to the proteolytic degradation of GHITM upon hyperpolarization of mitochondria (PubMed:35912435). Progressive GHITM degradation upon persistent hyperpolarization leads to respiratory complex I degradation and broad reshaping of the mitochondrial proteome by AFG3L2 (PubMed:35912435).

**Cellular Location**
Mitochondrion. Mitochondrion inner membrane {ECO:0000250|UniProtKB:Q8JZQ2}; Multi-pass membrane protein

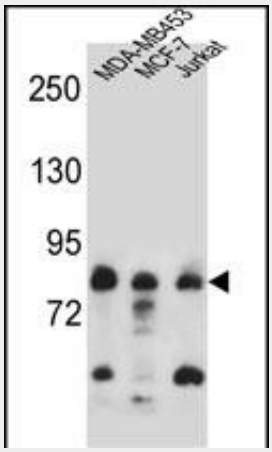
**Tissue Location**
Ubiquitous. Highly expressed in the cerebellar Purkinje cells.

## AFG3L2 Antibody (N-term) - Protocols

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

- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- 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).

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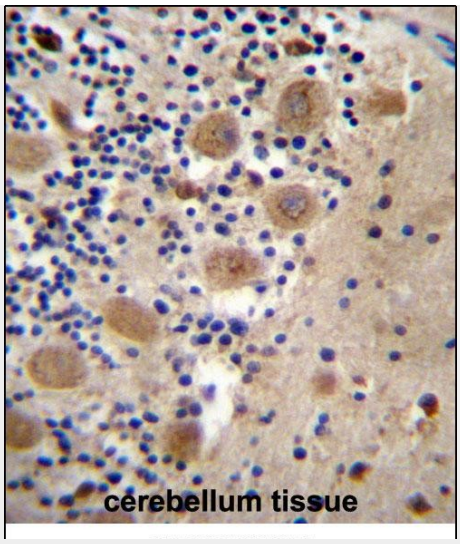


AFG3L2 Antibody (N-term) (Cat. #AP13219a)immunohistochemistry analysis in formalin fixed and paraffin embedded human cerebellum tissue followed by peroxidase conjugation of the secondary antibody and DAB staining.This data demonstrates the use of AFG3L2 Antibody (N-term) for immunohistochemistry. Clinical 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