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AFG3L2 Antibody (N-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP13219a

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
Clone Names	RB33011
Calculated MW	88584
Antigen Region	52-80

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~~1:100~500 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.

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.

Background

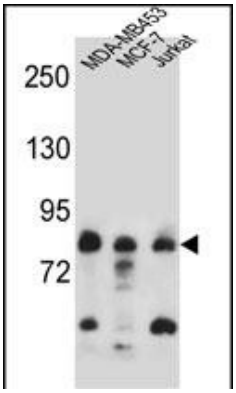
This gene encodes a protein localized in mitochondria and closely related to paraplegin. The paraplegin gene is responsible for an autosomal recessive form of hereditary spastic paraplegia. This gene is a candidate gene for other hereditary spastic paraplegias or neurodegenerative disorders.

References

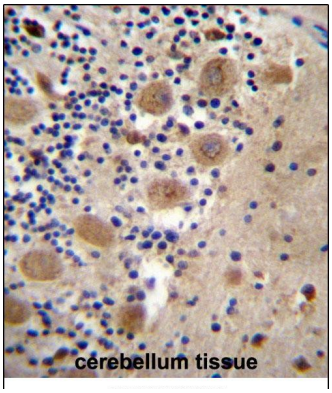
Edener, U., et al. Eur. J. Hum. Genet. 18(8):965-968(2010)
Di Bella, D., et al. Nat. Genet. 42(4):313-321(2010)
Augustin, S., et al. Mol. Cell 35(5):574-585(2009)
Mariotti, C., et al. Cerebellum 7(2):184-188(2008)
Cagnoli, C., et al. Brain 129 (PT 1), 235-242 (2006) :

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.

Citations

- Systematic analysis of a mitochondrial disease-causing ND6 mutation in mitochondrial deficiency