

AFG3L2 Polyclonal Antibody

Catalog # AP74280

Product Information

Application	WB, E
Primary Accession	Q9Y4W6
Reactivity	Human, Rat, Mouse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	88584

Additional Information

Gene ID	10939
Other Names	AFG3-like protein 2 (EC 3.4.24.-) (Paraplegin-like protein)
Dilution	WB~~WB 1:500-2000, ELISA 1:10000-20000 E~~N/A
Format	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.09% (W/V) sodium azide.
Storage Conditions	-20°C

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

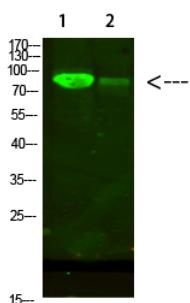
Tissue Location

Ubiquitous. Highly expressed in the cerebellar Purkinje cells.

Background

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 the maturation of paraplegin (SPG7) after its cleavage by mitochondrial-processing peptidase (MPP), converting it into a proteolytically active mature form (By similarity).

Images



Western Blot analysis of 1, mouse-heart 2,293T cells using primary antibody diluted at 1:500(4°C overnight). Secondary antibody : Goat Anti-rabbit IgG IRDye 800 (diluted at 1:5000, 25°C, 1 hour)

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