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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℃

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, PubMed:41075794). 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). Also acts as a regulator of carnitine biosynthesis by mediating cleavage and degradation of SLC25A45 (PubMed:41075794). 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).

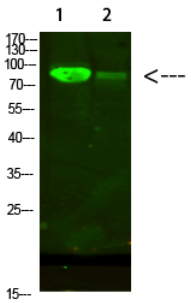
Cellular Location Mitochondrion inner membrane; Multi-pass membrane protein

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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