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Anti-AFG3L2 Picoband Antibody

Catalog # ABO12600

Product Information

Application	WB
Primary Accession	Q9Y4W6
Host	Rabbit
Reactivity	Human, Mouse, Rat
Clonality	Polyclonal
Format	Lyophilized
Description	Rabbit IgG polyclonal antibody for AFG3-like protein 2(AFG3L2) detection. Tested with WB in Human;Mouse;Rat.
Reconstitution	Add 0.2ml of distilled water will yield a concentration of 500ug/ml.

Additional Information

Gene ID	10939
Other Names	AFG3-like protein 2, 3.4.24.-, Paraplegin-like protein, AFG3L2
Calculated MW	88584
Application Details	Western blot, 0.1-0.5 □g/ml, Human, Mouse, Rat
Subcellular Localization	Mitochondrion membrane ; Multi-pass membrane protein .
Tissue Specificity	Ubiquitous. Highly expressed in the cerebellar Purkinje cells. .
Source	Eukaryota
Protein Name	AFG3-like protein 2
Contents	Each vial contains 5mg BSA, 0.9mg NaCl, 0.2mg Na2HPO4, 0.05mg NaN3.
Immunogen	E. coli-derived human AFG3L2 recombinant protein (Position: R168-D250). Human AFG3L2 shares 100% amino acid (aa) sequence identity with mouse AFG3L2.
Purification	Immunogen affinity purified.
Cross Reactivity	No cross reactivity with other proteins
Storage	At -20˚C for one year. After r˚Constitution, at 4˚C for one month. It˚Can also be aliquotted and stored frozen at -20˚C for a longer time.Avoid repeated freezing and thawing.

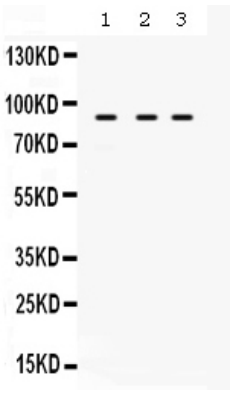
Protein Information

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

AFG3L2 is the catalytic subunit of the m-AAA protease, an ATP-dependent proteolytic complex of the mitochondrial inner membrane that degrades misfolded proteins and regulates ribosome assembly. In humans, it is encoded by the AFG3L2 gene. 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. And this gene is a candidate gene for other hereditary spastic paraplegias or neurodegenerative disorders as well as spastic ataxia-neuropathy syndrome.

Images



Western blot analysis of AFG3L2 expression in rat brain extract (lane 1), mouse spleen extract (lane 2) and U87 whole cell lysates (lane 3). AFG3L2 at 88KD was detected using rabbit anti- AFG3L2 Antigen Affinity purified polyclonal antibody (Catalog # ABO12600) at 0.5 ??g/mL. The blot was developed using chemiluminescence (ECL) method .

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