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SPG7 Antibody (Center) Blocking peptide
Synthetic peptide
Catalog # BP12476c

Specification

SPG7 Antibody (Center) Blocking peptide - Product Information

Primary Accession Q9UQ90

SPG7 Antibody (Center) Blocking peptide - Additional Information

Gene ID 6687

Other Names
Paraplegin, 3424-, Spastic paraplegia 7 protein, SPG7, CAR, CMAR, PGN

Format
Peptides are lyophilized in a solid powder format. Peptides can be reconstituted in solution using the appropriate buffer as needed.

Storage
Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C.

Precautions
This product is for research use only. Not for use in diagnostic or therapeutic procedures.

SPG7 Antibody (Center) Blocking peptide - Protein Information

Name SPG7 (HGNC:11237)

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:11549317, PubMed:28396416, PubMed:31097542, PubMed:9635427). SPG7 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 (By similarity). The m-AAA protease exerts a dual role in the mitochondrial inner membrane: it mediates the processing of specific regulatory proteins and ensures protein quality control by degrading misfolded polypeptides (By similarity). 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 (By similarity). 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:28396416, PubMed:31097542). Also

regulates mitochondrial calcium by catalyzing degradation of MCU (PubMed:31097542). Plays a role in the formation and regulation of the mitochondrial permeability transition pore (mPTP) and its proteolytic activity is dispensable for this function (PubMed:26387735).

Cellular Location
Mitochondrion inner membrane; Multi-pass membrane protein

Tissue Location
Ubiquitous.

SPG7 Antibody (Center) Blocking peptide - Protocols

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

- Blocking Peptides

SPG7 Antibody (Center) Blocking peptide - Images

SPG7 Antibody (Center) Blocking peptide - Background

This gene encodes a nuclear-encoded mitochondrialmetalloprotease protein that is a member of the AAA (ATPasesassociated with a variety of cellular activities) protein family.Members of this protein family share an ATPase domain and haveroles in diverse cellular processes including membrane trafficking,intracellular motility, organelle biogenesis, protein folding, andproteolysis. Two transcript variants encoding distinct isoformshave been identified for this gene. Mutations associated with thisgene cause autosomal recessive spastic paraplegia 7. [provided byRefSeq].

SPG7 Antibody (Center) Blocking peptide - References

Warnecke, T., et al. Mov. Disord. 25(4):413-420(2010)Augustin, S., et al. Mol. Cell 35(5):574-585(2009)Karlberg, T., et al. PLoS ONE 4 (10), E6975 (2009) :Brugman, F., et al. Neurology 71(19):1500-1505(2008)Tzoulis, C., et al. J. Neurol. 255(8):1142-1144(2008)