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SPG7 Antibody (Center)

Affinity Purified Rabbit Polyclonal Antibody (Pab)

Catalog # AP12476c

Specification

SPG7 Antibody (Center) - Product Information

Application	**IHC-P, WB,E**
Primary Accession	Q9UQ90
Other Accession	NP_955399.1, NP_003110.1
Reactivity	**Human**
Host	**Rabbit**
Clonality	**Polyclonal**
Isotype	**Rabbit IgG**
Calculated MW	**88235**
Antigen Region	**115-141**

SPG7 Antibody (Center) - Additional Information

Gene ID 6687

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

Target/Specificity
This SPG7 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 115-141 amino acids from the Central region of human SPG7.

Dilution
IHC-P~~1:10~50
WB~~1:1000
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
SPG7 Antibody (Center) is for research use only and not for use in diagnostic or therapeutic procedures.

SPG7 Antibody (Center) - 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

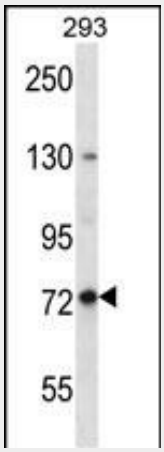
Tissue Location
Ubiquitous.

SPG7 Antibody (Center) - Protocols

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

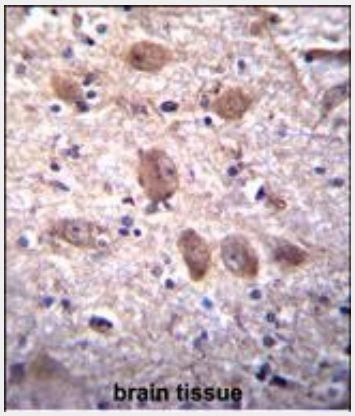
- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

SPG7 Antibody (Center) - Images



SPG7 Antibody (Center) (Cat. #AP12476c) western blot analysis in 293 cell line lysates

(35ug/lane).This demonstrates the SPG7 antibody detected the SPG7 protein (arrow).



SPG7 Antibody (Center) (Cat. #AP12476c)immunohistochemistry analysis in formalin fixed and paraffin embedded human brain tissue followed by peroxidase conjugation of the secondary antibody and DAB staining.This data demonstrates the use of SPG7 Antibody (Center) for immunohistochemistry. Clinical relevance has not been evaluated.

SPG7 Antibody (Center) - Background

This gene encodes a nuclear-encoded mitochondrial metalloprotease protein that is a member of the AAA (ATPases associated with a variety of cellular activities) protein family. Members of this protein family share an ATPase domain and have roles in diverse cellular processes including membrane trafficking, intracellular motility, organelle biogenesis, protein folding, and proteolysis. Two transcript variants encoding distinct isoforms have been identified for this gene. Mutations associated with this gene cause autosomal recessive spastic paraplegia 7. [provided by RefSeq].

SPG7 Antibody (Center) - 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)