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SPG3A Rabbit pAb

Catalog # AP97361

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q8WXF7
Other Accession	Q8WXF7
Reactivity	Rat, Mouse
Predicted	Human, Rabbit
Host	Rabbit
Clonality	Polyclonal
Calculated MW	63544
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human SPG3A/Atlastin
Epitope Specificity	201-300/558
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the GBP family. Atlastin subfamily.
SUBUNIT	Homooligomer. Interacts (via N-terminal region) withMAP4K4 (via CNH regulatory domain). Interacts with REEP5, RTN3 andRTN4 (via the transmembrane region). Interacts with SPAST;interaction is direct. May interact with TMED2. Interacts withREEP1.
DISEASE	Defects in ATL1 are the cause of spastic paraplegia autosomal dominant type 3 (SPG3) [MIM:182600]; also known as Strumpell-Lorrain syndrome. Spastic paraplegia is a degenerative spinal cord disorder characterized by a slow, gradual, progressive weakness and spasticity of the lower limbs.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Atlastins are Golgi-localized, integral membrane proteins that function as GTPases. The Atlastin proteins, also designated SPG3A and guanylate-binding protein 3, comprise a Dynamin superfamily that plays a role in axonal maintenance. Hereditary spastic paraplegia (HSP) is an inherited neurodegenerative disorder that is characterized by retrograde axonal degeneration. HSP primarily affects long corticospinal neurons and causes spastic lower extremity weakness. Spastin, a microtubule (MT)-severing AAA ATPase, is a binding partner of Atlastin that is involved in membrane dynamics. This Spastin/Atlastin binding may be involved in the biochemical pathway that leads to HSP development. Mutations in the Atlastin gene (SPG3A) account for approximately 10% of all autosomal dominant HSPs, while mutations in the Spastin gene (SPG4) account for almost 40%.

Additional Information

Gene ID	51062
Other Names	AD-FSP; ATL-1; FSP1; GBP3; HSN1D; SPG3; SPG3A; atlastin1; 4930435M24Rik;

	Adfsp; atlastin; ATLA1_BOVIN; ATL1; 3.6.5.-; ATLA1_HUMAN; Brain-specific GTP-binding protein; GTP-binding protein 3 (GBP-3 | hGBP3); Guanine nucleotide-binding protein 3; Spastic paraplegia 3 protein A; ATLA1_MOUSE; Spastic paraplegia 3A homolog; ATLA1_RAT; atlastin GTPase 1; spastic paraplegia 3A (autosomal dominant)
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	ATL1 (HGNC:11231)
Function	Atlastin-1 (ATL1) is a membrane-anchored GTPase that mediates the GTP-dependent fusion of endoplasmic reticulum (ER) membranes, maintaining the continuous ER network. It facilitates the formation of three-way junctions where ER tubules intersect (PubMed:14506257, PubMed:18270207, PubMed:19665976, PubMed:27619977, PubMed:34817557, PubMed:38509071). Two atlastin-1 on neighboring ER tubules bind GTP and form loose homodimers through the GB1/RHD3-type G domains and 3HB regions. Upon GTP hydrolysis, the 3HB regions tighten, pulling the membranes together to drive their fusion. After fusion, the homodimer disassembles upon release of inorganic phosphate (Pi). Subsequently, GDP dissociates, resetting the monomers to a conformation ready for a new fusion cycle (PubMed:14506257, PubMed:21220294, PubMed:21368113, PubMed:23334294, PubMed:38509071). May also regulate more or less directly Golgi biogenesis (PubMed:17321752). Indirectly regulates axonal development (By similarity).
Cellular Location	Endoplasmic reticulum membrane; Multi-pass membrane protein. Golgi apparatus membrane; Multi-pass membrane protein. Cell projection, axon {ECO:0000250|UniProtKB:Q6PST4}. Note=Localizes to endoplasmic reticulum tubular network (PubMed:27619977).
Tissue Location	Expressed predominantly in the adult and fetal central nervous system. Measurable expression in all tissues examined, although expression in adult brain is at least 50-fold higher than in other tissues. Detected predominantly in pyramidal neurons in the cerebral cortex and the hippocampus of the brain. Expressed in upper and lower motor neurons (at protein level)

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