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Rabbit Anti-SPG3A antibody

SL11759R

Product Name:	SPG3A
Chinese Name:	G蛋白Binding protein3抗体
Alias:	Atlastin; AD FSP; atl1; ATLA1_HUMAN; Atlastin GTPase 1; Atlastin-1; Atlastin1; Brain specific GTP binding protein; Brain-specific GTP-binding protein; FSP1; GBP-3; GBP3; GTP-binding protein 3; Guanine nucleotide-binding protein 3; Guanylate binding protein 3; hGBP3; HSN1D; Spastic paraplegia 3 protein A; SPG 3A; SPG3; SPG3A.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Rabbit,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	64kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human SPG3A/Atlastin:201-300/558
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	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%.

Function:
GTPase tethering membranes through formation of trans-homooligomer and mediating homotypic fusion of endoplasmic reticulum membranes. Functions in endoplasmic reticulum tubular network biogenesis. May also regulate Golgi biogenesis. May regulate axonal development.

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.

Subcellular Location:
Endoplasmic reticulum membrane. Golgi apparatus membrane. Cell projection > axon.

Tissue Specificity:
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).

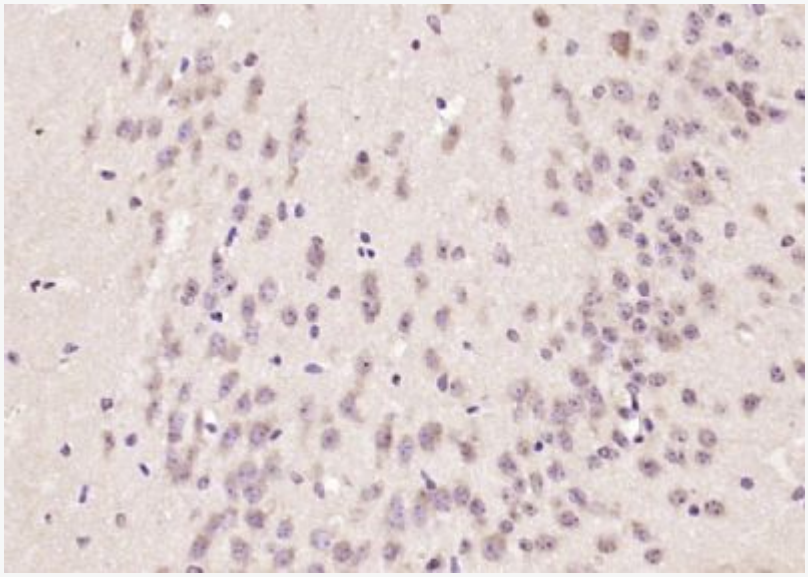
DISEASE:
Defects in ATL1 are the cause of spastic paraplegia autosomal dominant type 3 (SPG3) [MIM:182600]; also known as Strumpell-Lorrain syndrome. Spastic paraplegia is a degenerative spinal cord disorder characterized by a slow, gradual, progressive weakness and spasticity of the lower limbs.

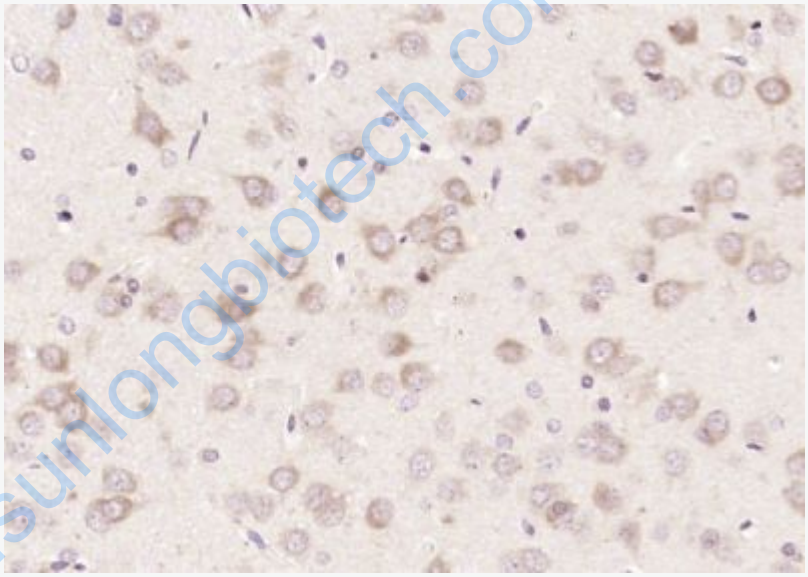
Similarity:
Belongs to the GBP family. Atlastin subfamily.

SWISS:
Q8WXF7

Gene ID:
51062

Database links:

	Entrez Gene: 51062 Human Entrez Gene: 73991 Mouse Entrez Gene: 362750 Rat Entrez Gene: 535424 Cow Omim: 606439 Human SwissProt: Q58D72 Cow SwissProt: Q8WXF7 Human SwissProt: Q8BH66 Mouse SwissProt: Q6PST4 Rat Unigene: 584905 Human Unigene: 474462 Mouse Unigene: 135117 Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	

	Paraformaldehyde-fixed, paraffin embedded (mouse brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (SPG3A) Polyclonal Antibody, Unconjugated (SL11759R) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.
	
	Paraformaldehyde-fixed, paraffin embedded (rat brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (SPG3A) Polyclonal Antibody, Unconjugated (SL11759R) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.