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Rabbit Anti-SPG21 antibody

SL11785R

Product Name:	SPG21
Chinese Name:	痉挛性截瘫相关蛋白21抗体
Alias:	Acid cluster protein 33; ACP33; BM019; BM-019; GL010; MAST; Maspardin; Spastic paraplegia 21 autosomal recessive Mast syndrome protein; SPG21 antibody; SPG21_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Pig,Horse,Rabbit,Sheep,
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:	35kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human SPG21:151-250/308
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:	Maspardin is a 308 amino acid cytoplasmic protein that is widely expressed. Belonging to the AB hydrolase superfamily, Maspardin colocalizes with CD4 on endosomal/trans-Golgi network. It is thought that Maspardin may act as a negative regulatory factor in CD4-dependent T-cell activation. Defects in the gene encoding Maspardin are the result of hereditary spastic paraplegia autosomal recessive type 21 (also designated Mast

	syndrome), an autosomal recessive neurodegenerative disorder characterized by a slow, gradual, progressive weakness and spasticity of the lower limbs. The gene encoding Maspardin is encoded by human chromosome 15, which houses over 700 genes and comprises nearly 3% of the human genome. **Function:** Defects in SPG21 are the cause of Mast syndrome, an autosomal recessive hereditary spastic paraplegia with dementia and other CNS abnormalities (SPG21). Present at high frequency among the Old Order Amish. Subtle childhood abnormalities may be present, but the main features develop in early adulthood. The disease is slowly progressive, and cerebellar and extrapyramidal signs are also found in patients with advanced disease. Patients have a thin corpus callosum and white matter abnormalities. The protein encoded by this gene was identified by a two hybrid screen using CD4 as the bait. It binds to the hydrophobic C terminal amino acids of CD4 which are involved in repression of T cell activation. The interaction with CD4 is mediated by the noncatalytic alpha/beta hydrolase fold domain of this protein. It is thus proposed that this gene product modulates the stimulatory activity of CD4. **Subunit:** Interacts with CD4. Interacts with ALDH16A1. **Subcellular Location:** Cytoplasm; cytosol. Membrane; peripheral membrane protein. **Tissue Specificity:** Expressed in all tissues tested, including heart, brain, placenta, lung, liver, skeletal muscle, kidney and pancreas. Expressed in J.CaM1.6, HuT 78 and HeLa cell lines (at protein level). **DISEASE:** Defects in SPG21 are the cause of spastic paraplegia autosomal recessive type 21 (SPG21) [MIM:248900]; also known as Mast syndrome. Spastic paraplegia is a neurodegenerative disorder characterized by a slow, gradual, progressive weakness and spasticity of the lower limbs. Rate of progression and the severity of symptoms are quite variable. Initial symptoms may include difficulty with balance, weakness and stiffness in the legs, muscle spasms, and dragging the toes when walking. In some forms of the disorder, bladder symptoms (such as incontinence) may appear, or the weakness and stiffness may spread to other parts of the body. SPG21 is associated with dementia and other central nervous system abnormalities. Subtle childhood abnormalities may be present, but the main features develop in early adulthood. The disease is slowly progressive, and cerebellar and extrapyramidal signs are also found in patients with advanced disease. Patients have a thin corpus callosum and white-matter abnormalities. **Similarity:** Belongs to the AB hydrolase superfamily.

SWISS:
Q9NZD8

Gene ID:
51324

Database links:

Entrez Gene: 404069Cow

Entrez Gene: 101866571Cynomolgus monkey

Entrez Gene: 51324Human

Entrez Gene: 27965Mouse

Entrez Gene: 100174603Orangutan

NCBI: NP_057714.1Human

Omim: 608181Human

SwissProt: Q8MJJ1Cow

SwissProt: Q4R5H6Cynomolgus monkey

SwissProt: Q9NZD8Human

SwissProt: Q9CQC8Mouse

SwissProt: Q5RES2Orangutan

Unigene: 242458Human

Important Note:
This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.