内质网脂质转运相关蛋白 2 抗体

产品货号： mlR10720

英文名称： SPFH2

中文名称： 内质网脂质转运相关蛋白 2 抗体

别　　名： C8orf2; Chromosome 8 open reading frame 2; Endoplasmic reticulum lipid raft associated protein 2; Endoplasmic reticulum lipid raft-associated protein 2; ER lipid raft associated 2; ERLIN 2; Erlin-2; ERLIN2; ERLN2_HUMAN; HGNC:1356; MGC87072; NET32; SPFH 2; SPFH domain containing protein 2; SPFH domain family member 2; SPFH domain-containing protein 2; SPFH2; SPG18; Stomatin prohibitin flotillin HflC/K domain containing protein 2; Stomatin-prohibitin-flotillin-HflC/K domain-containing protein 2.

研究领域： 细胞生物　神经生物学　信号转导　表观遗传学

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Pig, Cow, Rabbit, Sheep,

产品应用： WB=1:500-2000 ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500（石蜡切片需做抗原修复）
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

分 子 量： 38kDa

细胞定位： 细胞浆 细胞膜

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human SPFH2:231-330/339

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： 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

产品介绍： SPFH2 is a ubiquitously expressed 339 amino acid protein that belongs to the band 7/mec-2 family. Localized to lipid raft-like domains in the membrane of the endoplasmic reticulum (ER), SPFH2 plays a crucial role in the ER-associated degradation (ERAD) pathway that removes metabolically regulated and aberrant proteins from the ER. Specifically, SPFH2 associates with IP3R-I (Inositol 1,4,5-triphosphate receptor I), a substrate of the ERAD pathway, and facilitates its polyubiquitination and subsequent degradation. Three isoforms of SPFH2 are expressed due to alternative splicing events.

Function:

Component of the ERLIN1/ERLIN2 complex which mediates the endoplasmic reticulum-associated degradation (ERAD) of inositol 1,4,5-trisphosphate receptors (IP3Rs). Also involved in ITPR1 degradation by the ERAD pathway.

Subunit:

Interacts with activated ITPR1, independently of the degree of ITPR1 polyubiquitination (By similarity). Forms a heteromeric complex with ERLIN1. In complex with ERLIN1, interacts with RNF170.

Subcellular Location:

Endoplasmic reticulum membrane. Associated with lipid raft-like domains of the endoplasmic reticulum membrane.

Tissue Specificity:

Ubiquitous.

DISEASE:

Defects in ERLIN2 are the cause of spastic paraplegia autosomal recessive type 18 (SPG18) [MIM:611225]. A form of spastic paraplegia, 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. SPG18 is a severe form with onset in early childhood. Most affected individuals have severe psychomotor retardation. Some may develop significant joint contractures.

Similarity:

Belongs to the band 7/mec-2 family.

SWISS:

O94905

Gene ID:

11160

Important Note:

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