α 肌萎缩糖蛋白 2 抗体

产品货号： mlR8740

英文名称： alpha Sarcoglycan

中文名称： α 肌萎缩糖蛋白 2 抗体

别　　名： 50 DAG; 50 kDa dystrophin associated glycoprotein; 50 kDa dystrophin-associated glycoprotein; 50DAG; 50kD DAG; 59kDa; A2; adhalin; ADL; Alpha SG; Alpha-sarcoglycan; Alpha-SG; Asg; DAG2; DMDA2; Dystroglycan 2; Dystroglycan-2; LGMD2D; sarcoglycan, alpha (dystrophin-associated glycoprotein); SCARMD1; Sgca; SGCA_HUMAN.

研究领域： 细胞生物　神经生物学　信号转导　糖蛋白

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Dog, Pig, Rabbit,

产品应用： WB=1:500-2000 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： 40kDa

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 alpha Sarcoglycan:51-150/387 <Extracellular>

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： Preservative: 15mM Sodium Azide, Constituents: 1% BSA, 0.01M PBS, pH 7.4

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 background:

Component of the sarcoglycan complex, a subcomplex of the dystrophin-glycoprotein complex which forms a link between the F-actin cytoskeleton and the extracellular matrix.

Function:

Component of the sarcoglycan complex, a subcomplex of the dystrophin-glycoprotein complex which forms a link between the F-actin cytoskeleton and the extracellular matrix.

Subunit:

Interacts with the syntrophin SNTA1. Cross-link to form 2 major subcomplexes: one consisting of SGCB, SGCD and SGCG and the other consisting of SGCB and SGCD. The association between SGCB and SGCG is particularly strong while SGCA is loosely associated with the other sarcoglycans (By similarity).

Subcellular Location:

Cell membrane > sarcolemma. Cytoplasm > cytoskeleton.

Tissue Specificity:

Most strongly expressed in skeletal muscle. Also expressed in cardiac muscle and, at much lower levels, in lung. In the fetus, most abundant in cardiac muscle and, at lower levels, in lung. Also detected in liver and kidney. Not expressed in brain.

DISEASE:

Defects in SGCA are the cause of limb-girdle muscular dystrophy type 2D (LGMD2D) [MIM:608099]; also known as Duchenne-like muscular dystrophy autosomal recessive type 2 or severe childhood autosomal recessive muscular dystrophy (SCARMD). LGMD2D is an autosomal recessive degenerative myopathy characterized by progressive muscle wasting from early childhood with loss of independent ambulation by teenage years. Muscle

biopsy shows necrosis, decreased immunostaining for alpha sarcoglycan, and adhalin deficiency. The phenotype is less severe than LGMD2C.

Similarity:

Belongs to the sarcoglycan alpha/epsilon family.

SWISS:

Q16586

Gene ID:

6442

Important Note:

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

产品图片

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