

产品名称: Dysferlin Rabbit Polyclonal Antibody

产品货号: APRab10243

产品概述 (Summary)

描述(Description)	Rabbit polyclonal Antibody
宿主(Host)	Rabbit
应用(Application)	WB,ICC/IF,ELISA
种属反应(Reactivity)	Human,Mouse
偶联物(Conjugation)	Unconjugated
修饰(Modification)	Unmodified
同种型(Isotype)	IgG
克隆(Clonality)	Polyclonal
剂型(Form)	Liquid
保存条件(Storage)	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
储存溶液(Buffer)	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
纯化 (Purification)	Affinity purification

产品应用(Application)

稀释比(Dilution Ratio)	WB 1:500-1:2000,ICC/IF 1:200-1:1000,ELISA 1:5000-1:20000
分子量(Molecular Weight)	240kDa

抗原信息(Antigen Information)

基因名(Gene Name)	DYSF
别名(Alternative Names)	DYSF; FER1L1; Dysferlin; Dystrophy-associated fer-1-like protein; Fer-1-like protein 1
基因 ID(Gene ID)	8291.0
SwissProt ID	O75923
免疫原(Immunogen)	The antiserum was produced against synthesized peptide derived from human Dysferlin. AA range:1981-2030

研究背景 (Background)

dysferlin(DYSF) Homo sapiens The protein encoded by this gene belongs to the ferlin family and is a skeletal muscle protein found associated with the sarcolemma. It is involved in muscle contraction and contains C2 domains that play a role in calcium-

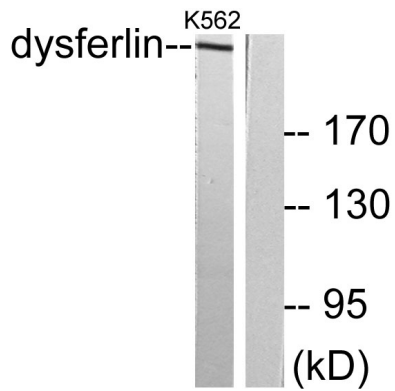
mediated membrane fusion events, suggesting that it may be involved in membrane regeneration and repair. In addition, the protein encoded by this gene binds caveolin-3, a skeletal muscle membrane protein which is important in the formation of caveolae. Specific mutations in this gene have been shown to cause autosomal recessive limb girdle muscular dystrophy type 2B (LGMD2B) as well as Miyoshi myopathy. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Aug 2008],developmental stage:Expression in limb tissue from 5-6 weeks embryos; persists throughout development.,disease:Defects in DYSF are the cause of distal myopathy with anterior tibial onset (DMAT) [MIM:606768]. Onset of the disorder is between 14 and 28 years of age and the anterior tibial muscles are the first muscle group to be involved. Inheritance is autosomal recessive.,disease:Defects in DYSF are the cause of limb-girdle muscular dystrophy type 2B (LGMD2B) [MIM:253601]. LGMD2B is an autosomal recessive degenerative myopathy characterized by weakness and atrophy starting in the proximal pelvifemoral muscles, with onset in the late teens or later, massive elevation of serum creatine kinase levels and slow progression. Scapular muscle involvement is minor and not present at onset. Upper limb girdle involvement follows some years after the onset in lower limbs.,disease:Defects in DYSF are the cause of Miyoshi myopathy (MM) [MIM:254130]. This type of autosomal recessive muscular dystrophy involves the distal lower limb musculature. It is characterized by weakness that initially affects the gastrocnemius muscle during early adulthood. Otherwise the phenotype overlaps with LGMD2B, especially in age at onset and creatine kinase elevation.,domain:The C2 domain 1 associates with lipid membranes in a calcium-dependent manner.,function:Key calcium ion sensor involved in the $\text{Ca}(2+)$ -triggered synaptic vesicle-plasma membrane fusion. Plays a role in the sarcolemma repair mechanism of both skeletal muscle and cardiomyocytes that permits rapid resealing of membranes disrupted by mechanical stress.,online information:Dysferlin,online information:Dysferlin entry,sequence caution:Translation N-terminally shortened.,similarity:Belongs to the ferlin family.,similarity:Contains 5 C2 domains.,subcellular location:Colocalizes, during muscle differentiation, with BIN1 in the T-tubule system of myotubules and at the site of contact between two myotubes or a myoblast and a myotube. Wounding of myotubes led to its focal enrichment to the site of injury and to its relocation in a $\text{Ca}(2+)$ -dependent manner toward the plasma membrane. Colocalizes with AHNAK, AHNAK2 and PARVB at the sarcolemma of skeletal muscle. Detected on the apical plasma membrane of the syncytiotrophoblast. Reaches the plasmma membrane through a caveolin-independent mechanism. Retained by caveolin at the plasmma membrane (By similarity). Colocalizes, during muscle differentiation, with CACNA1S in the T-tubule system of myotubules (By similarity). Accumulates and colocalizes with fusion vesicles at the sarcolemma disruption sites.,subunit:Interacts with CACNA1S. Interacts with ANXA1; the interaction is $\text{Ca}(2+)$ - and injury state-dependent. Interacts with ANXA2; the interaction is $\text{Ca}(2+)$ - and injury state-dependent (By similarity). Interacts with CAV3 and PARVB. Interacts with AHNAK; the interaction is direct and $\text{Ca}(2+)$ -independent. Interacts with AHNAK2; the interaction is direct and $\text{Ca}(2+)$ -independent.,tissue specificity:Expressed in skeletal muscle, myoblast, myotube and in the syncytiotrophoblast (STB) of the placenta (at protein level). Highly expressed in skeletal muscle. Also found in heart, brain, spleen, intestine, placenta and at lower levels in liver, lung, kidney and pancreas.,

研究领域 (Research Area)

注意事项 (Note)

For research use only.

图片 (Image Data)



Western blot analysis of lysates from K562 cells, using Dysferlin Antibody. The lane on the right is blocked with the synthesized peptide.

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产品类别	产品货号	产品名称
WB 解决方案	RA10020	2h 极速 WB 即用型全流程试剂盒
	RA10021	4h 快速 WB 即用型全流程试剂盒
	RA10042	5h 畅享版 WB 全流程试剂盒
	RA10037	校准级彩色预染蛋白 Marker (8-180kDa)
	RA10038	校准级彩色预染蛋白 Marker (10-250kDa)
	RA10039	校准级高分子彩色预染蛋白 Marker (25-400kDa)
TSA 多重荧光染色试剂盒	RA10008	TSA 双标三色多重荧光染色试剂盒 (mIHC)
	RA10009	TSA 三标四色多重荧光染色试剂盒 (mIHC)
	RA10010	TSA 四标五色多重荧光染色试剂盒 (mIHC)
	RA10011	TSA 五标六色多重荧光染色试剂盒 (mIHC)
	RA10012	TSA 六标七色多重荧光染色试剂盒 (mIHC)
IHC 检测试剂盒	RA10006	HRP Anti-Mouse/Rabbit IHC Detection System
	RA10007	Polymer-HRP Anti-Mouse/Rabbit IHC Detection System
抗体标记试剂盒	RE80004p	辣根过氧化物酶(HRP)抗体标记试剂盒
	RE80002q	Sulfo-NHS-生物素标记试剂盒
	RE80007p	Cy3 荧光素标记试剂盒
	RE80011p	Fluor488 荧光素标记试剂盒
	RE80017p	Fluor750 荧光素标记试剂盒
	RE80005p	藻红蛋白(R-PE) 抗体快速标记试剂盒
	RE80040	PE-Cy7 串联染料抗体快速标记试剂盒
稳转细胞系构建服务 (免费赠送全膜 WB 验证)	TS-0001	过表达稳转细胞系构建
	TS-0002	敲低稳转细胞系构建
	TS-0003	敲除细胞系构建

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