

Summary

Production Name	Dysferlin Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	WB
Reactivity	Human,Mouse

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

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

Application

Dilution Ratio	WB 1:500-1:2000. ELISA: 1:10000.
Molecular Weight	240kD

Background

dysferlin(DYSF) Homo sapiens The protein encoded by this gene belongs to the ferlin family and is a skeletal muscle

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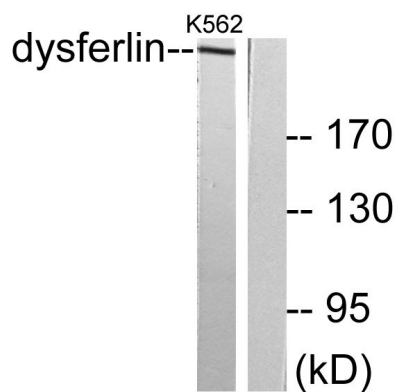
protein found associated with the sarcolemma. It is involved in muscle contraction and contains C2 domains that play a role in calcium-mediated membrane fusion events, suggesting that it may be involved in membrane regeneration and repair. In addition, the protein encoded by this gene binds caveolin-3, 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 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 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 plasma membrane through a caveolin-independent mechanism. Retained by caveolin at the plasma 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 Ca(2+)- and injury state-dependent. Interacts with ANXA2; the interaction is Ca(2+)- and injury state-dependent (By similarity). Interacts with CAV3 and PARVB. Interacts with AHNAK; the interaction is direct and Ca(2+)-independent. Interacts with AHNAK2; the interaction is direct and 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.,

Research Area

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Image Data



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

Note

For research use only.