SunLong Biotech Co.,LTD
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[Primary Antibody]

Rabbit Anti-Dysferlin interacting protein 1 antibody

Catalog Number: SL14476R

FOR RESEARCH USE ONLY

[DATASHEET]

Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	Dysferlin interacting protein 1
Immunogen:	KLH conjugated synthetic peptide derived from human Dysferlin interacting protein 1: 7-100/154
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Applications:	WB=1:500-2000 （Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Reactivity:	Mouse,Rat(predicted:Human,Dog,Cow,Horse,Sheep)
Predicted MW.:	17kDa
Background:	Dysferlin is a muscle-specific protein that is essential for normal muscle function and development (1). Mutations in the human dysferlin gene, DYSF, which maps to chromosome 2p13.3-p13.1, are associated with limb girdle muscular dystrophy-2B (LGMD-2B) and a related, adult-onset, distal dystrophy known as Miyoshi myopathy (MM) (1,2). Dysferlin localizes to the muscle fiber membrane, but is absent in MM and LGMD-2B muscle (1,3). Dysferlin is detected in 5-6 week embryos, when limbs begin to form regional differentiation (3). Although it is not essential for initial myogenesis, dysferlin appears to be critical for sustained normal function in mature muscle (1). It has been suggested that the absence of dysferlin during development gives rise to the disease phenotype in adulthood (3). Identical mutations in the dysferlin gene can produce more than one myopathy phenotype, indicating that additional genes and/or other factors are also involved in the clinical phenotype (4,5). The DYSF gene has no homology to any other known mammalian gene, but the protein product is related to the

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	spermatogenesis factor fer-1 of Caenorhabditis elegans. The name 'dysferlin' combines the role of the gene in producing muscular dystrophy with its homology to C. elegans (6). **Function:** Inhibits phosphatase activity of protein phosphatase 1 (PP1) complexes. **Subunit:** Interacts with DYSF and PPP1CA. **Similarity:** Contains 2 ANK repeats. **Database links:** **Entrez Gene:** 116729 Human **Entrez Gene:** 616223 Cow **Entrez Gene:** 608003 Dog **Entrez Gene:** 68701 Mouse **Entrez Gene:** 287881 Rat **SwissProt:** Q86WC6 Human **SwissProt:** Q9D119 Mouse

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