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Dysferlin interacting protein 1 Rabbit pAb

Catalog # AP96592

Product Information

Application	WB
Primary Accession	Q86WC6
Other Accession	Q86WC6
Reactivity	Rat, Mouse
Predicted	Human, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	17438
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Dysferlin interacting protein 1
Epitope Specificity	7-100/154
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 2 ANK repeats.
SUBUNIT	Interacts with DYSF and PPP1CA.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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 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).

Additional Information

Gene ID	116729
Other Names	DYSFIP1; 1110033I14Rik; toonin; RGD1308861; PPR27_HUMAN; PPP1R27;

	Dysferlin-interacting protein 1; PPR27_MOUSE; protein phosphatase 1 regulatory subunit 27; dysferlin-interacting protein 1 (toonin); dysferlin interacting protein 1 (toonin); dysferlin interacting protein 1; protein phosphatase 1, regulatory subunit 27
Dilution	WB=1:500-2000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	PPP1R27
Synonyms	DYSFIP1
Function	Inhibits phosphatase activity of protein phosphatase 1 (PP1) complexes.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.