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Product datasheet for AP06090PU-N

Dysferlin (DYSF) Rabbit Polyclonal Antibody

Product data:

Product Type:	Primary Antibodies
Applications:	ELISA, IF, WB
Recommended Dilution:	**Western blot**: 1/500-1/1000. **Immunofluorescence**: 1/50-1/200.
Reactivity:	Human, Mouse
Host:	Rabbit
Clonality:	Polyclonal
Specificity:	This antibody detects endogenous levels of DP-1/TFDP1 protein (region surrounding Pro2013).
Formulation:	Phosphate buffered saline (PBS), pH 7.2. State: Aff - Purified State: Liquid purified Ig fraction Preservative: 0.05% Sodium azide
Concentration:	1.0 mg/ml
Purification:	Affinity chromatography using epitope-specific immunogen (> 95% pure by SDS-PAGE).
Conjugation:	Unconjugated
Storage:	Store undiluted at 2-8°C for one month or (in aliquots) at -20°C for longer. Avoid repeated freezing and thawing.
Stability:	Shelf life: one year from despatch.
Predicted Protein Size:	~240 kDa
Gene Name:	dysferlin
Database Link:	Entrez Gene 26903 MouseEntrez Gene 8291 Human O75923

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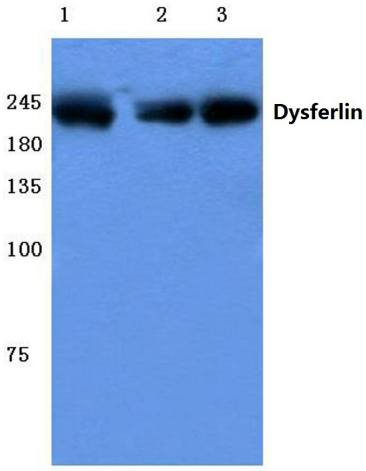
Background: Dysferlin is a muscle-specific protein that is essential for normal muscle function and development. Mutations in the human dysferlin gene, DYSF, which maps to chromosome 2p13.3-p13.1, are associated with limb girdle muscular dystrophy-2B (LGMD-2B) and a related, adult-onset, distal dystrophy known as Miyoshi myopathy (MM).
Dysferlin, a protein with a molecular mass of approximately 230 kDa, localizes to the muscle fiber membrane, but is absent in MM and LGMD-2B muscle. Dysferlin is detected in 5-6 week embryos, when limbs begin to form regional differentiation. Although it is not essential for initial myogenesis, dysferlin appears to be critical for sustained normal function in mature muscle. It has been suggested that the absence of dysferlin during development gives rise to the disease phenotype in adulthood. 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.
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.

Synonyms: Fer-1-like protein 1, DYSF, FER1L1

Protein Families: Transmembrane

Product images:



Western blot (WB) analysis of Dysferlin antibody (Cat.-No.: AP06090PU-N) at 1/500 dilution Lane 1:MCF-7 cell lysate Lane 2:sp2/0 cell lysate Lane 3:Rat liver tissue lysate