

Dysferlin (P2013) polyclonal antibody

Catalog: BS1094

Host: Rabbit

Reactivity: Human, Mouse, Rat

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*.

Product:

1 mg/ml in Phosphate buffered saline (PBS) with 0.05% sodium azide, approx. pH 7.2.

Molecular Weight:

~ 240 kDa

Swiss-Prot:

O75923

Purification&Purity:

The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen and the purity is > 95% (by SDS-PAGE).

Applications:

WB: 1:500~1:1000

IF: 1:50~1:200

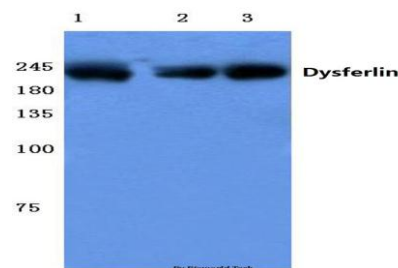
Storage&Stability:

Store at 4 °C short term. Aliquot and store at -20 °C long term. Avoid freeze-thaw cycles.

Specificity:

Dysferlin (P2013) pAb detects endogenous levels of DP-1/TFDP1 protein.

DATA:



Western blot (WB) analysis of Dysferlin (P2013) pAb at 1:500 dilution

Lane1:MCF-7 cell lysate

Lane2:sp2/0 cell lysate

Lane3:Rat liver tissue lysate

Note:

For research use only, not for use in diagnostic procedure.

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