Bioworld Technology CO., Ltd.

Dysferlin (P2013) Peptide

Cat No.: BS1094P

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 nvolved 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.

Swiss-Prot

O75923

Applications

Blocking

Specificity

This peptide can be used with studies using BS1094 Dysferlin (P2013) pAb.

Purification & Purity

Synthetic peptide Dysferlin (P2013). (Note: the amino acid sequence is proprietary). The purity is > 98%.

Product

1 mg/ml in DI water.

Storage & Stability

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

Research Use

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

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