bs-14476R **[Primary Antibody]**

Dysferlin interacting protein 1 Rabbit pAb

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DATASHEET

Host: Rabbit **Isotype:** IgG

Clonality: Polyclonal

GeneID: 116729 **SWISS:** Q86WC6

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°C. Store at -20°C for one year. Avoid repeated freeze/thaw cycles.

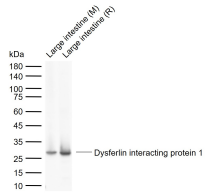
Background: Dysferlin is a muscle-specific protein that is essential for normal muscle function and development (1). Mutations in the human dysferlin gene, DYSF, which maps to chromosome 2p13.3-p13.1, are associated with limb girdle muscular dystrophy-2B (LGMD-2B) and a related, adult-onset, distal dystrophy 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 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).

Applications: **WB** (1:500-2000)

Reactivity: Mouse, Rat (predicted: Human, Sheep, Cow, Dog, Horse)

Predicted MW.: 17 kDa

VALIDATION IMAGES



Sample: Lane 1: Mouse Large intestine tissue lysates Lane 2: Rat Large intestine tissue lysates Primary: Anti-Dysferlin interacting protein 1 (bs-14476R) at 1/1000 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 17 kDa Observed band size: 27 kDa