Dysferlin Conjugated Antibody

Catalog No: #C49359

Package Size: #C49359-AF350 100ul #C49359-AF405 100ul #C49359-AF488 100ul #C49359-AF555 100ul #C49359-AF594 100ul #C49359-AF647 100ul #C49359-AF680 100ul #C49359-AF750 100ul #C49359-Biotin 100ul

Orders: order@signalwayantibody.com
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Description

Product Name	Dysferlin Conjugated Antibody
Host Species	Rabbit
Clonality	Monoclonal
Species Reactivity	Hu, Ms
Immunogen Description	recombinant protein
Conjugates	Biotin AF350 AF405 AF488 AF555 AF594 AF647 AF680 AF750
Other Names	DMAT antibody DYSF antibody DYSF_HUMAN antibody Dysferlin antibody Dysferlin limb girdle muscular dystrophy 2B (autosomal recessive) antibody Dysferlin limb girdle muscular dystrophy 2B antibody Dystrophy associated fer 1 like 1 antibody Dystrophy associated fer 1 like protein antibody Dystrophy associated fer1 like 1 antibody Dystrophy associated fer1 like protein antibody Dystrophy-associated fer-1-like protein antibody Fer 1 like protein 1 antibody Fer-1-like protein 1 antibody Fer1 like protein 1 antibody FER1L1 antibody FLJ00175 antibody FLJ90168 antibody LGMD 2B antibody LGMD2B antibody Limb girdle muscular dystrophy 2B (autosomal recessive) antibody Limb girdle muscular dystrophy 2B antibody Miyoshi myopathy antibody MM antibody MMD1 antibody
Accession No.	Swiss-Prot#:O75923
Uniprot	O75923
GeneID	8291;
Excitation Emission	AF350: 346nm/442nm AF405: 401nm/421nm AF488: 493nm/519nm AF555: 555nm/565nm AF594: 591nm/614nm AF647: 651nm/667nm AF680: 679nm/702nm AF750: 749nm/775nm
Calculated MW	237 kDa
Formulation	0.01M Sodium Phosphate, 0.25M NaCl, pH 7.6, 5mg/ml Bovine Serum Albumin, 0.02% Sodium Azide
Storage	Store at 4°C in dark for 6 months

Application Details

Suggested Dilution:

AF350 conjugated: most applications: 1: 50 - 1: 250

AF405 conjugated: most applications: 1: 50 - 1: 250

AF488 conjugated: most applications: 1: 50 - 1: 250

AF555 conjugated: most applications: 1: 50 - 1: 250

AF594 conjugated: most applications: 1: 50 - 1: 250

AF647 conjugated: most applications: 1: 50 - 1: 250

AF680 conjugated: most applications: 1: 50 - 1: 250

AF750 conjugated: most applications: 1: 50 - 1: 250

Biotin conjugated: working with enzyme-conjugated streptavidin, most applications: 1: 50 - 1: 1,000

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 2p13.2, are associated with limb girdle muscular dystrophy-2B (LGMD-2B) and a related, adult-onset, distal dystrophy known as Miyoshi myopathy (MM). Dysferlin 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 oΩ½oΩ½dysferlinoΩ½oΩ½ combines the role of the gene in producing muscular dystrophy with its homology to C. elegans.

Note: This product is for in vitro research use only