Anti-Dysferlin antibody

Description	Rabbit polyclonal to Dysferlin.

Model	STJ92803
Host	Rabbit
Reactivity	Human, Mouse
Applications	ELISA, IF, WB
Immunogen	Synthesized peptide derived from human Dysferlin
Immunogen Region	1950-2030 aa, C-terminal
Gene ID	8291
Gene Symbol	DYSF
Dilution range	WB 1:500-1:2000IF 1:200-1:1000ELISA 1:10000
Specificity	Dysferlin Polyclonal Antibody detects endogenous levels of Dysferlin protein.
Tissue Specificity	Expressed in skeletal muscle, myoblast, myotube and in the syncytiotrophoblast (STB) of the placenta (at protein level). Ubiquitous. Highly expressed in skeletal muscle. Also found in heart, brain, spleen, intestine, placenta and at lower levels in liver, lung, kidney and pancreas.
Purification	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.
Note	For Research Use Only (RUO).
Protein Name	Dysferlin Dystrophy-associated fer-1-like protein Fer-1-like protein 1
Molecular Weight	240 kDa

Clonality	Polyclonal
Conjugation	Unconjugated
Isotype	IgG
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Concentration	1 mg/ml
Storage Instruction	Store at -20°C, and avoid repeat freeze-thaw cycles.
Database Links	HGNC:3097OMIM:253601
Alternative Names	Dysferlin Dystrophy-associated fer-1-like protein Fer-1-like protein 1
Function	Key calcium ion sensor involved in the Ca(2+)-triggered synaptic vesicle-plasma membrane fusion. Plays a role in the sarcolemma repair mechanism of both skeletal muscle and cardiomyocytes that permits rapid resealing of membranes disrupted by mechanical stress .
Sequence and Domain Family	All seven C2 domains associate with lipid membranes in a calcium-dependent manner. Domains C2 1 and 3 have the highest affinity for calcium, the C2 domain 1 seems to be largely unstructured in the absence of bound ligands. The C2 domain 1 from isoform 14 does not bind calcium in the absence of bound phospholipid .
Cellular Localization	Cell membrane, sarcolemma. Single-pass type II membrane protein. Cytoplasmic vesicle membrane Cell membrane. Colocalizes, during muscle differentiation, with BIN1 in the T-tubule system of myotubules and at the site of contact between two myotubes or a myoblast and a myotube. Wounding of myotubes led to its focal enrichment to the site of injury and to its relocalization in a Ca(2+)-dependent manner toward the plasma membrane. Colocalizes with AHNAK, AHNAK2 and PARVB at the sarcolemma of skeletal muscle. Detected on the apical plasma membrane of the syncytiotrophoblast. Reaches the plasmma membrane through a caveolin-independent mechanism. Retained by caveolin at the plasmma membrane . Colocalizes, during muscle differentiation, with CACNA1S in the T-tubule system of myotubules . Accumulates and colocalizes with fusion vesicles at the sarcolemma disruption sites .