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Anti-Dysferlin Antibody

Catalog # AP51171

Product Information

Application	WB, IF/IC
Primary Accession	O75923
Reactivity	Human, Mouse, Rat, Pig, Bovine, Dog
Host	Rabbit
Clonality	Polyclonal
Calculated MW	237295

Additional Information

Gene ID	8291
Other Names	FER1L1; Dysferlin; Dystrophy-associated fer-1-like protein; Fer-1-like protein 1
Target/Specificity	KLH-conjugated synthetic peptide encompassing a sequence within the C-term region of human Dysferlin. The exact sequence is proprietary.
Dilution	WB~~WB (1/500 - 1/1000) IF/IC~~N/A
Format	Liquid in 0.42% Potassium phosphate, 0.87% Sodium chloride, pH 7.3, 30% glycerol, and 0.01% sodium azide.
Storage	Store at -20 °C.Stable for 12 months from date of receipt

Protein Information

Name	DYSF
Synonyms	FER1L1
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 (By similarity).
Cellular Location	Cell membrane, sarcolemma; Single-pass type IV membrane protein. Cytoplasmic vesicle membrane {ECO:0000250|UniProtKB:Q9ESD7}; Single-pass type IV membrane protein. Cell membrane; Single-pass type IV membrane protein. Late endosome membrane; Single-pass type IV membrane protein. Note=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 (By similarity). Colocalizes, during muscle differentiation, with CACNA1S in the T-tubule system of myotubules (By similarity). Accumulates and colocalizes with fusion vesicles at the sarcolemma disruption sites (By similarity) {ECO:0000250|UniProtKB:Q9ESD7}

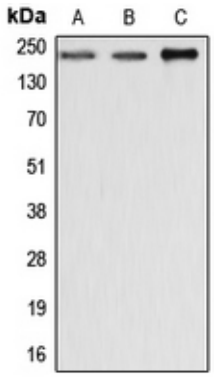
Tissue Location

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.

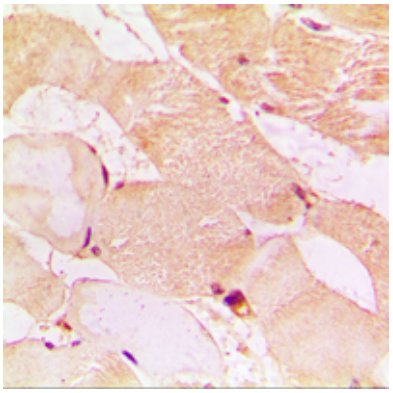
References

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Hillier L.W.,et al.Nature 434:724-731(2005).
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Images

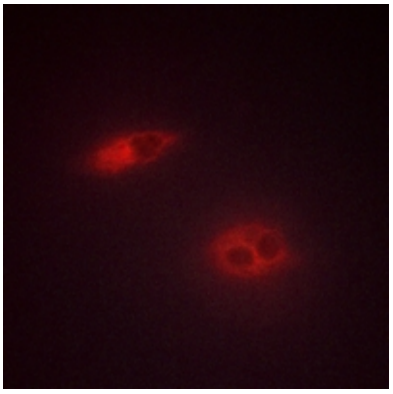


Western blot analysis of Dysferlin expression in MCF7 (A), SP2/0 (B), rat liver (C) whole cell lysates. (Predicted band size: 237 kD; Observed band size: 237 kD)



Immunohistochemical analysis of Dysferlin staining in human muscle formalin fixed paraffin embedded tissue section. The section was pre-treated using heat mediated antigen retrieval with sodium citrate buffer (pH 6.0). The section was then incubated with the antibody at room temperature and detected using an HRP conjugated compact polymer system. DAB was used as the chromogen. The section was then counterstained with haematoxylin and mounted with DPX.

Immunofluorescent analysis of Dysferlin staining in HeLa cells. Formalin-fixed cells were permeabilized with 0.1% Triton X-100 in TBS for 5-10 minutes and blocked with 3% BSA-PBS for 30 minutes at room temperature. Cells were probed with the primary antibody in 3% BSA-PBS and incubated overnight at 4 °C in a hidified chamber. Cells were washed with PBST and incubated with a



AF594-conjugated secondary antibody (red) in PBS at room temperature in the dark.

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