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Product Information

Anti-Dysferlin (N-terminal region)
produced in rabbit, affinity isolated antibody

Catalog Number **SAB4200388**

Product Description

Anti-Dysferlin (N-terminal region) is produced in rabbit using as immunogen a synthetic peptide corresponding to a sequence in the N-terminal region of human dysferlin (GeneID: 8291), conjugated to KLH. The corresponding sequence is identical in human dysferlin isoforms 1-14 and highly conserved (84% sequence identity) in rat and mouse dysferlin. The antibody is affinity-purified using the immunizing peptide immobilized on agarose.

Anti-Dysferlin (N-terminal region) specifically recognizes human and mouse dysferlin. The antibody may be used in several immunochemical techniques including immunoblotting (~250 kDa), immunoprecipitation and immunofluorescence. Detection of the dysferlin band by immunoblotting is specifically inhibited by the dysferlin immunizing peptide.

Muscular dystrophy (MD) includes a diverse group of inherited muscle diseases characterized by slow progressive weakness and loss of skeletal muscle.[1] More than 30 gene loci have been identified that cause MD emphasizing the heterogeneity of the disease. Mutations in the dysferlin gene DYSF cause limb-girdle muscle dystrophy type 2B (LGMD2B), an autosomal recessive disorder, and the related Miyoshi myopathy.[1,2] Dysferlin is a transmembrane protein that belongs to the ferlin-1 family of proteins including myoferlin and otoferlin and is homologous to the c. elegans fer-1 protein.[1,2] Dysferlin is expressed early during human development and has been implicated in membrane fusion events. It has been suggested to play a role in membrane repair processes, such as the ability to reseal the sarcolemma upon muscle injury.[2,3] The integral membrane proteins caveolin-1 and -3 have been shown to regulate the endocytosis of dysferlin.[4] Dysferlin localization in the membrane and trafficking is impaired by mutations in caveolin-1 and -3, resulting in mis-targeting and redistribution of dysferlin from the plasma membrane to the Golgi complex.[4,5]

Reagent

Supplied as a solution in 0.01 M phosphate buffered saline, pH 7.4, containing 15 mM sodium azide.

Antibody Concentration: ~1.5 mg/mL

Precautions and Disclaimer

This product is for R&D use only, not for drug, household, or other uses. Please consult the Material Safety Data Sheet for information regarding hazards and safe handling practices.

Storage/Stability

For continuous use, store at 2-8 °C for up to one month. For extended storage, freeze in working aliquots. Repeated freezing and thawing, or storage in "frost-free" freezers, is not recommended. If slight turbidity occurs upon prolonged storage, clarify the solution by centrifugation before use. Working dilutions should be discarded if not used within 12 hours.

Product Profile

Immunoblotting: a working concentration of 0.5-1.0 µg/mL is recommended using HEK-293T cells over-expressing human dysferlin.

Immunoprecipitation: a working amount of 5-10 µg is recommended using HEK-293T cells over-expressing human dysferlin.

Immunofluorescence: a working concentration of 7-14 µg/mL is recommended using differentiated C2C12 myoblasts.

Note: In order to obtain the best results using various techniques and preparations, we recommend determining the optimal working dilutions by titration.

References

1. Laval, S.H., and Bushby, K.M., *Neuropathl. Appl. Neurobiol.,* **30**, 91-105 (2004).
2. Glover, L., and Brown, R.H., *Traffic*, **8**, 785-794 (2007).
3. Bansal, D., et al., *Nature,* **423**, 168-172 (2003).
4. Hernandez-Deviez, D.J., et al., *Hum. Mol. Genet*., **15**, 129-142 (2006).
5. Hernandez-Deviez, D.J., et al., *J. Biol. Chem.,* **283**, 6476-6488 (2008).

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