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Product Information

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

Catalog Number **SAB4200453**

Product Description

Anti-Dysferlin (N-terminal) is developed 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 identical in mouse dysferlin. The antibody is affinity-purified using the immunizing peptide immobilized on agarose.

Anti-Dysferlin (N-terminal) specifically recognizes human, rat, and mouse dysferlin. The antibody may be used in several immunochemical techniques including immunoblotting (~250 kDa), immunofluorescence, and immunohistochemistry. 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 mistargeting 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: ~1.0 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 is not recommended. Storage in “frost-free” freezers is also 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 0.1–0.2 μg/mL is recommended using extracts of HEK-293T cells overexpressing human dysferlin and 1–2 μg/mL using A10 cells.

Immunofluorescence: a working concentration of 5–10 μg/mL is recommended using differentiated C2C12 myoblasts.

Immunohistochemistry: a working concentration of 20–30 μg/mL is recommended using methanol-acetone fixed frozen sections of mouse skeletal muscle.

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).