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Product Information

Anti-Dystrophin antibody, Mouse monoclonal
clone MANDRA1, purified from hybridoma cell culture

Product Number **SAB4200763**

Product Description

Anti-Dystrophin antibody, Mouse monoclonal (mouse IgG1 isotype) is derived from the MANDRA1 hybridoma produced by the fusion of mouse myeloma cells and splenocytes from BALB/c mice immunized with a fusion protein containing the C-terminal fragment of human dystrophin (GeneID: 1756).[1-3] The isotype is determined by ELISA using Mouse Monoclonal Antibody Isotyping Reagents, Product Number ISO2. The antibody is purified from culture supernatant of hybridoma cells.

Anti-Dystrophin antibody, Mouse monoclonal specifically recognizes an epitope located on the 128 amino acids at the end of the C-terminal domain of the human dystrophin molecule [2,4,5] The antibody reacts with dystrophin from human,[1,2,5,6] mouse,[2] rat,[5] and zebrafish[7] origin. Dystrophin is expressed in striated muscles from nearly all Becker muscular dystrophies, but is absent in cases of Duchenne muscular dystrophies and in the dystrophic mouse (mdx).[1-3,13] The antibody is specific to dystrophin and does not react with actinin or utrophin, an autosomal homologue of dystrophin that is also known as dystrophin-related protein (DRP).[2,5] The antibody may be used in various immunochemical techniques including immunoblotting (~427 kDa),[8] immuno-histochemistry,[9] ELISA,[10] and immunofluorescence.[11] The antibody also recognizes the 70-75 kDa protein, now known as apo-dystrophin-1 or DP71[1,4] which is detected in the brain[5] as well as in lympho blastoid cells, cultures of brain astroglial and neuronal cells, liver, and Hep G2 cells (human hepatoma).[4,5]

Dystrophin is a rod-shaped cytoskeletal protein located to the periphery (plasma membrane) of normal striated muscle fibers. Dystrophin is absent, reduced, or altered as a result of mutation in Duchenne and Becker muscular dystrophies (DMD/BMD) or in its homologue in mouse.[12,13] Severe DMD is associated with a marked dystrophin deficiency whereas patients with the milder form of BMD show less pronounced abnormalities of protein expression.

Anti-Dystrophin antibody, Mouse monoclonal provides means for studying dystrophin protein structure and function, interactions with other proteins and the nature of the partial gene products produced in some patients carrying deletions in the dystrophin gene. The antibody may be useful in the prenatal or post-abortion diagnosis of muscular dystrophy carriers by immunohistological analyses.

Reagent

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

Antibody Concentration: ~1.0 mg/mL

Precautions and Disclaimer

For R&D use only. Not for drug, household, or other uses. Please consult the 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. If slight turbidity occurs upon prolonged storage, clarify the solution by centrifugation before use. Working dilution samples should be discarded if not used within 12 hours.

Product Profile

Immunohistochemistry: a working concentration of 10–20 µg/mL is recommended using acetone fixed rat tongue frozen sections.

Note: In order to obtain best results in various techniques and preparations, it is recommended to determine optimal working dilutions by titration.

References

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