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ProductInformation

MONOCLONAL ANTI-DYSTROPHIN
Clone MANDYS8
Mouse Ascites Fluid

Product Number **D 8168**

Product Description

Monoclonal Anti-Dystrophin (mouse IgG2b isotype) is derived from the MANDYS8 hybridoma produced by the fusion of mouse myeloma cells and splenocytes from an immunized mouse. A recombinant human dystrophin fragment was used as the immunogen. The isotype is determined using Sigma ImmunoType™ Kit (Product Code ISO-1) and by a double diffusion immunoassay using Mouse Monoclonal Antibody Isotyping Reagents (Product Code ISO-2).

Monoclonal Anti-Dystrophin, clone MANDYS8, recognizes an epitope located on the rod domain of the human dystrophin molecule.[1,2] This epitope is present in normal muscle tissue and in nearly all Becker muscular dystrophies. It is absent in cases of Duchenne muscular dystrophies and in the dystrophic mouse (mdx). Immunohistochemical staining of muscle tissue with the antibody results in clear labeling, confined to the periphery (plasma membrane) of normal striated muscle fibers. The epitope recognized by the antibody is sensitive to formalin fixation and paraffin embedding. The antibody exhibits a wide interspecies cross-reactivity (e.g., human, mouse, rat, chicken, rabbit, porcine). It may also be used in immunoblotting of brain dystrophin and in ELISA.

Dystrophin is a muscle membrane protein (427 kDa), which is absent, reduced, or altered as a result of mutation in the Duchenne and Becker muscular dystrophies (DMD/BMD) gene, or its homologue in the mouse.[3] Severe DMD is associated with a marked dystrophin deficiency, while patients with the milder form of BMD show less pronounced abnormalities of protein expression. Since abnormalities in the protein expression occur specifically in patients with these types of muscular dystrophy, dystrophin analysis may be used to distinguish these conditions from other neuromuscular diseases. Predictions from the sequence suggest a structural protein on the inner face of the membrane, consisting of a 25-repeat, rod-like, triple-helical domain separating an N-terminal actin-binding domain from two C-terminal domains, one of which is rich in cysteine.[4]

The large size of dystrophin and its low abundance (<0.01% of the total muscle protein) hinder isolation of intact, native protein for structure/function studies. Monoclonal antibodies against defined regions of dystrophin provide a means of studying its 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. They are also useful in the prenatal or post-abortion diagnosis of muscle dystrophies carriers by immunohistological analyses.[5]

Monoclonal Anti-Dystrophin, clone MANDYS8, may be used for the localization of dystrophin using various immunochemical assays such as ELISA, immunoblot, and immunohistochemistry.

Reagents

The product is provided as ascites fluid with 0.1% sodium azide as a preservative.

Precautions and Disclaimer

Due to the sodium azide content a material safety data sheet (MSDS) for this product has been sent to the attention of the safety officer of your institution. Consult the MSDS for information regarding hazards and safe handling practices.

Storage/Stability

For continuous use, store at 2-8 °C for a maximum of one month. For extended storage, solution may be frozen in working aliquots. Repeated freezing and thawing is not recommended. If slight turbidity occurs upon prolonged storage, clarify by centrifugation before use.

Product Profile

The minimum working dilution is 1:400 by indirect immunofluorescent labeling of freshly dissected or frozen human or animal muscle tissue.

In order to obtain best results, it is recommended that each individual user determine their optimal working dilution by titration assay.

References
1. Nguyen, thi Man, et al., FEBS Lett., **262**, 237 (1990).
2. Sedgwick, S., et al., Nucleic Acids Res., **19**, 5889 (1991).
3. Hoffman, E., et al., Cell, **51**, 919 (1987).
4. Koenig, M., et al., Cell, **53**, 219 (1988).
5. Ginjaar, I., et al., Lancet, **338**, 258 (1991).

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