



3050 Spruce Street  
Saint Louis, Missouri 63103 USA  
Telephone 800-325-5832 • (314) 771-5765  
Fax (314) 286-7828  
email: techserv@sial.com  
sigma-aldrich.com

## Product Information

### Monoclonal Anti-Dystrophin Clone MANDRA1 Mouse Ascites Fluid

Product Number **D 8043**

#### Product Description

Monoclonal Anti-Dystrophin (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 fusion protein containing the C-terminal 485 amino acids (3200-3684) of human dystrophin.<sup>1-3</sup> 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 MANDRA1, recognizes an epitope located on the 128 amino acids at the end of the C-terminal domain of the human dystrophin molecule (a.a. residues 3558-3684).<sup>2,4,5</sup> This epitope is present in normal striated muscle. It 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).<sup>1-3</sup> The antibody is specific to dystrophin and does not react with  $\alpha$ -actinin or utrophin, an autosomal homologue of dystrophin that is also known as dystrophin-related protein (DRP).<sup>2,5</sup> Immunohistochemical staining of muscle tissue with the antibody results in labeling confined to the periphery (plasma membrane) of normal striated muscle fibers.<sup>1,2,6</sup> The antibody stains dystrophin (427 kDa) in muscle<sup>1,2,5</sup> and brain<sup>5</sup> extracts in immunoblotting. It recognizes the 70-75 kDa protein, now known as apo-dystrophin-1 or DP71,<sup>4</sup> which is detected in the brain<sup>5</sup> as well as in lympho blastoid cells, cultures of brain astroglial and neuronal cells, liver and Hep G2 cells (human hepatoma).<sup>4,5</sup> 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,<sup>1,2,5,6</sup> mouse,<sup>2,7</sup> rat,<sup>5</sup> and fish<sup>2</sup>). It may be used in ELISA<sup>2</sup> and in capture ELISA.<sup>7</sup>

Monoclonal Anti-Dystrophin, clone MANDRA1, may be used for the localization of dystrophin using various

immunochemical assays such as ELISA, capture ELISA, immunoblot, and immunohistochemistry.

Dystrophin is a muscle membrane protein (427 kDa) which is absent, reduced or altered as a result of mutation in Duchenne and Becker muscular dystrophies (DMD/BMD) or its homologue in the mouse.<sup>8</sup> Severe DMD is associated with a marked dystrophin deficiency whereas patients with the milder form of BMD show less pronounced abnormalities of protein expression. Because 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.<sup>9</sup> The large size of dystrophin and its low abundance (<0.01% of the total muscle protein) are a hindrance to the isolation of intact, native protein for structure/function studies. Monoclonal antibodies against defined regions<sup>10</sup> of dystrophin provide a means for 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. The antibodies are useful in the prenatal or post-abortion diagnosis of muscular dystrophy carriers by immunohistological analyses.<sup>11</sup>

#### 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 sheet (MSDS) for this product has been sent to the attention of the safety officer of your institution. Consult the MSDS for information regarding hazardous 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 not recommended. If slight turbidity occurs upon prolonged storage, clarify the solution by centrifugation before use.

### **Product Profile**

The minimum antibody titer of 1:100 was determined by indirect immunofluorescent staining of freshly dissected and frozen human or animal muscle tissue. In order to obtain best results, it is recommended that each user determine the optimal working dilution for individual applications by titration assay.

### **References**

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PCS 08/02

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