

# Product datasheet

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# ARG43809 anti-DMD / Dystrophin antibody [DMD/6270]

Package: 50 μg Store at: -20°C

#### **Summary**

Host

Clone

Product Description Mouse Monoclonal antibody [DMD/6270] recognizes DMD / Dystrophin

Tested Reactivity Hu
Tested Application IHC-P

**Clonality** Monoclonal

Isotype IgG2a

Target Name DMD / Dystrophin

Species Human

Immunogen Synthetic peptide corresponding to a.a 1700-2300 within Spectrin like repeats region of Human DMD /

Dystrophin.

Mouse

DMD/6270

Conjugation Un-conjugated

Alternate Names BMD; CMD3B; DXS270; DXS272; MRX85; Dystrophin; DXS164; DXS239; DXS206; DXS142; DXS230;

DXS269; DXS268

## **Application Instructions**

Application table	Application	Dilution
	IHC-P	1 - 2 μg/ml
Application Note	IHC-P: Antigen retrieval: boil tissue sections in 10 mM Tris with 1 mM EDTA (pH 9.0) for 20 min and allow samples to cool to RT before staining.  * The dilutions indicate recommended starting dilutions and the optimal dilutions or concentrations should be determined by the scientist.	
Positive Control	Human heart tissue	

## **Properties**

Form Liquid

Purification Purification with Protein A/G affinity.

Buffer PBS, 0.05% Sodium azide and 0.1 mg/ml BSA.

Preservative 0.05% Sodium azide

Stabilizer 0.1 mg/ml BSA

Concentration 0.2 mg/ml

Storage instruction For continuous use, store undiluted antibody at 2-8°C for up to a week. For long-term storage, aliquot

and store at -20°C or below. Storage in frost free freezers is not recommended. Avoid repeated

freeze/thaw cycles. Suggest spin the vial prior to opening. The antibody solution should be gently mixed before use.

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For laboratory research only, not for drug, diagnostic or other use.

#### Bioinformation

Gene Symbol

Note

DMD

Gene Full Name

dystrophin

Background

The dystrophin gene is the largest gene found in nature, measuring 2.4 Mb. The gene was identified through a positional cloning approach, targeted at the isolation of the gene responsible for Duchenne (DMD) and Becker (BMD) Muscular Dystrophies. DMD is a recessive, fatal, X-linked disorder occurring at a frequency of about 1 in 3,500 new-born males. BMD is a milder allelic form. In general, DMD patients carry mutations which cause premature translation termination (nonsense or frame shift mutations), while in BMD patients dystrophin is reduced either in molecular weight (derived from in-frame deletions) or in expression level. The dystrophin gene is highly complex, containing at least eight independent, tissue-specific promoters and two polyA-addition sites. Furthermore, dystrophin RNA is differentially spliced, producing a range of different transcripts, encoding a large set of protein isoforms. Dystrophin (as encoded by the Dp427 transcripts) is a large, rod-like cytoskeletal protein which is found at the inner surface of muscle fibers. Dystrophin is part of the dystrophin-glycoprotein complex (DGC), which bridges the inner cytoskeleton (F-actin) and the extra-cellular matrix. [provided by RefSeq, Jul 2008]

Function

Anchors the extracellular matrix to the cytoskeleton via F-actin. Ligand for dystroglycan. Component of the dystrophin-associated glycoprotein complex which accumulates at the neuromuscular junction (NMJ) and at a variety of synapses in the peripheral and central nervous systems and has a structural function in stabilizing the sarcolemma. Also implicated in signaling events and synaptic transmission. [UniProt]

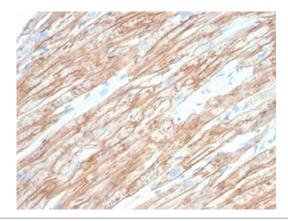
Calculated Mw

427 kDa

**Cellular Localization** 

Cell membrane, sarcolemma; Peripheral membrane protein; Cytoplasmic side. Cytoplasm, cytoskeleton. Cell junction, synapse, postsynaptic cell membrane. Note=In muscle cells, sarcolemma localization requires the presence of ANK2, while localization to costameres requires the presence of ANK3. Localizes to neuromuscular junctions (NMJs). In adult muscle, NMJ localization depends upon ANK2 presence, but not in newborn animals. [UniProt]

#### **Images**



ARG43809 anti-DMD / Dystrophin antibody [DMD/6270] IHC-P image

Immunohistochemistry: Paraffin-embedded Human heart tissue was stained with ARG43809 anti-DMD / Dystrophin antibody [DMD/6270]. Antigen Retrieval: Boil tissue section in buffer contains 10 mM Tris and 1 mM EDTA (pH 9.0) for 20 min.