

# Product datasheet

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# ARG41594 anti-DMD / Dystrophin antibody

Package: 100 μl Store at: -20°C

#### **Summary**

Product Description Rabbit Polyclonal antibody recognizes DMD / Dystrophin

Tested Reactivity Hu, Ms, Rat

Tested Application ICC/IF, IHC-P, WB

Host Rabbit

Clonality Polyclonal

Isotype IgG

Target Name DMD / Dystrophin

Species Human

Immunogen Recombinant fusion protein corresponding to aa. 346-635 within Spectrin like repeats domain of

Human DMD / Dystrophin (NP\_004007.1).

Conjugation Un-conjugated

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

DXS269; DXS268

### **Application Instructions**

Application table	Application	Dilution
	ICC/IF	1:50 - 1:200
	IHC-P	1:50 - 1:200
	WB	1:500 - 1:2000
Application Note	* The dilutions indicate recommended starting dilutions and the optimal dilutions or concentrations should be determined by the scientist.	

#### **Properties**

Form Liquid

Purification Affinity purified.

Buffer PBS (pH 7.3), 0.02% Sodium azide and 50% Glycerol.

Preservative 0.02% Sodium azide

Stabilizer 50% Glycerol

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. 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.

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

#### Bioinformation

Gene Symbol

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]

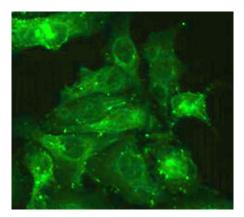
Calculated Mw

427 kDa

**Cellular Localization** 

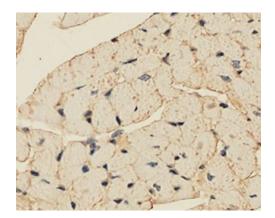
Cell membrane, sarcolemma; Peripheral membrane protein; Cytoplasmic side. Cytoplasm, cytoskeleton. Cell junction, synapse, postsynaptic cell membrane. Note=In muscle cells, sarcolemma localization requires the presence of ANK2, while localization to costameres requires the presence of ANK3. Localizes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

#### **Images**



#### ARG41594 anti-DMD / Dystrophin antibody ICC/IF image

Immunofluorescence: HeLa cells stained with ARG41594 anti-DMD / Dystrophin antibody at  $1:100\ dilution$ .



## ARG41594 anti-DMD / Dystrophin antibody IHC-P image

Immunohistochemistry: Paraffin-embedded Mouse heart tissue stained with ARG41594 anti-DMD / Dystrophin antibody at 1:100 dilution.