

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

Package: 50 µg
Store at: -20°C

Summary

Product Description	Mouse Monoclonal antibody [DMD/6270] recognizes DMD / Dystrophin
Tested Reactivity	Hu
Tested Application	IHC-P
Host	Mouse
Clonality	Monoclonal
Clone	DMD/6270
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.
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

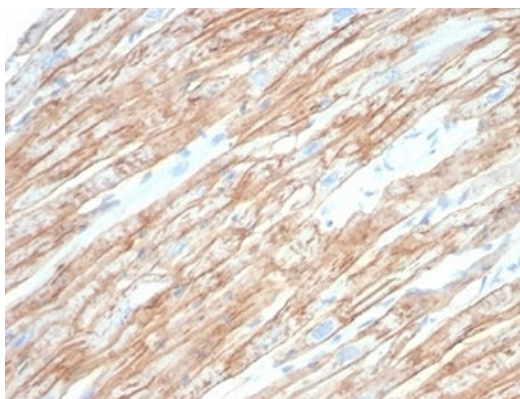
freeze/thaw cycles. Suggest spin the vial prior to opening. The antibody solution should be gently mixed before use.

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



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.