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DATA SHEET

Mouse Anti-Dystrophin (N-terminal Actin Binding Domain) [DMD/3243]: MC0340, MC0340RTU7

Intended Use: For Research Use Only

Description: Dystrophin is a rod-shaped protein, measuring about 150 nm, consisting of 3684 amino acids with a calculated molecular weight of 427 kDa. Dystrophin can be separated into four domains: N-terminal actin binding domain (amino acids 14-240), central rod domain (amino acids 253-3040), Cysteine-rich domain (amino acids 3080-3360), and C-terminal domain (amino acids 3361-3685). Its N-terminal domain binds to F-Actin cytoskeleton on the inner surface of muscle fibers to the surrounding extracellular matrix, through the cell membrane interface. Its C-terminal domain binds to the dystrophin-associated glycoprotein (DAG) complex in the membrane. The human dystrophin gene measures 2.4 megabases, has more than 80 exons, produces a 14 kb mRNA and contains at least 8 independent tissue-specific promoters and 2 poly A sites. The dystrophin mRNA can undergo differential splicing and produce a range of transcripts that encode a large set of proteins. Dystrophin represents approximately 0.002% of total striated muscle protein and localizes to triadic junctions in skeletal muscle, where it is thought to influence calcium ion homeostasis and force transmission. Pathogenic mutations in dystrophin result in Duchenne (DMD) and Becker (BMD) muscular dystrophies. This clone is recommended for detection of an epitope corresponding to amino acids 114-263 of dystrophin N-terminal actin binding domain.

Specifications

Clone:	DMD/3243
Source:	Mouse
Isotype:	IgG1k
Reactivity:	Human
Immunogen:	Recombinant human dystrophin fragment aa 114-263
Localization:	Membrane and cytoplasm
Formulation:	Purified antibody in PBS pH7.4, containing BSA and ≤ 0.09% sodium azide (NaN3)
Storage:	Store at 2°- 8°C
Applications:	IHC, ELISA

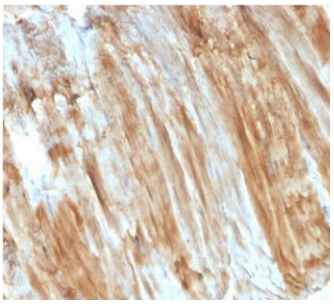
Package:

Description	Catalog No.	Size
Dystrophin (N-terminal Actin Binding Domain) Concentrated	MC0340	1 ml
Dystrophin (N-terminal Actin Binding Domain) Prediluted	MC0340RTU7	7 ml

IHC Procedure*

Positive Control Tissue:	Skeletal muscle and heart muscle tissues
Concentrated Dilution:	50-200
Pretreatment:	Tris EDTA pH9.0, 15 minutes Pressure Cooker or 30-60 minutes water bath at 95°-99°C
Incubation Time and Temp:	30-60 minutes @ RT
Detection:	Refer to the detection system manual

* Result should be confirmed by an established diagnostic procedure.



FFPE human skeletal muscle stained with anti-Dystrophin using DAB

References:

1. CRISPR/Cas9-generated mouse model of Duchenne muscular dystrophy recapitulating a newly identified large 430 kb deletion in the human DMD gene. Egorova TV, et al. Dis Model Mech 12:N/A, 2019.
2. Use of capillary Western immunoassay (Wes) for quantification of dystrophin levels in skeletal muscle of healthy controls and individuals with Becker and Duchenne muscular dystrophy. Beekman C, et al. PLoS One 13:e0195850, 2018.

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