

**Mouse Anti-Dystrophin [DMD/3243]: MC0340, MC0340RTU7**

**Intended Use:** For Research Use Only

**Description:** Dystrophin-glycoprotein complex (DGC) connects the F-Actin cytoskeleton on the inner surface of muscle fibers to the surrounding extracellular matrix, through the cell membrane interface. A deficiency in this protein contributes to Duchenne (DMD) and Becker (BMD) muscular dystrophies. 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.

**Specifications**

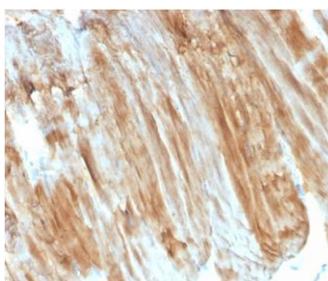
Clone: DMD/3243  
 Source: Mouse  
 Isotype: IgG1k  
 Reactivity: Human  
 Immunogen: OTN 11, ovarian carcinoma cell line  
 Localization: Membrane and cytoplasm  
 Formulation: Antibody in PBS pH7.4, containing BSA and  $\leq 0.09\%$  sodium azide (NaN<sub>3</sub>)  
 Storage: Store at 2°- 8°C  
 Applications: IHC, ELISA  
 Package:

Description	Catalog No.	Size
Dystrophin [DMD/3243] Concentrated	MC0340	1 ml
Dystrophin [DMD/3243] 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 using Pressure Cooker, or 30-60 minutes using 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.
3. Function and genetics of dystrophin and dystrophin related proteins in muscle. Blake DJ, et al. Physiol Rev 82: 291-329, 2002.

Doc. 100-MC0340  
Rev. A