

dystrophin (H-300): sc-15376

BACKGROUND

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.

CHROMOSOMAL LOCATION

Genetic locus: DMD (human) mapping to Xp21.2; Dmd (mouse) mapping to X B.

SOURCE

dystrophin (H-300) is a rabbit polyclonal antibody raised against amino acids 801-1100 mapping within an internal region of dystrophin of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

dystrophin (H-300) is recommended for detection of dystrophin of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

dystrophin (H-300) is also recommended for detection of dystrophin in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for dystrophin siRNA (h): sc-35240, dystrophin siRNA (m): sc-35241, dystrophin shRNA Plasmid (h): sc-35240-SH, dystrophin shRNA Plasmid (m): sc-35241-SH, dystrophin shRNA (h) Lentiviral Particles: sc-35240-V and dystrophin shRNA (m) Lentiviral Particles: sc-35241-V.

Molecular Weight of dystrophin: 427 kDa.

Positive Controls: L8 Cell Lysate : sc-3807 or A-10 Cell Lysate : sc-3806.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

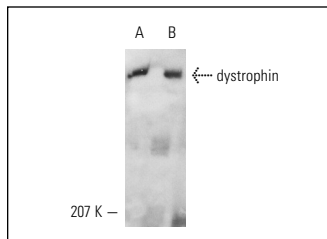
PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.

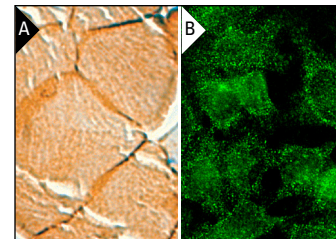
STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

DATA



dystrophin (H-300): sc-15376. Western blot analysis of dystrophin expression in L8 (A) and A-10 (B) whole cell lysates



dystrophin (H-300): sc-15376. Immunoperoxidase staining of formalin fixed, paraffin-embedded smooth muscle tissue showing membrane and cytoplasmic localization (A). Immunofluorescence staining of methanol-fixed HeLa cells showing cytoskeletal localization (B).

SELECT PRODUCT CITATIONS

- Rodríguez, M., et al. 2005. Ischemia depletes dystrophin and inhibits protein synthesis in the canine heart: mechanisms of myocardial ischemic injury. *J. Mol. Cell. Cardiol.* 38: 723-733.
- Ozasa, S., et al. 2007. Efficient conversion of ES cells into myogenic lineage using the gene-inducible system. *Biochem. Biophys. Res. Commun.* 357: 957-963.
- Xiong, D., et al. 2007. Inducible cardiac-restricted expression of enteroviral protease 2A is sufficient to induce dilated cardiomyopathy. *Circulation* 115: 94-102.
- Zhang, J., et al. 2008. Syncoilin is required for generating maximum isometric stress in skeletal muscle but dispensable for muscle cytoarchitecture. *Am. J. Physiol., Cell Physiol.* 294: C1175-C1182.
- Noé, L., et al. 2010. Platelet G_s hypofunction and abnormal morphology resulting from a heterozygous RGS2 mutation. *J. Thromb. Haemost.* 8: 1594-1603.
- Boulberda, M., et al. 2011. Genetic inactivation of prokineticin receptor-1 leads to heart and kidney disorders. *Arterioscler. Thromb. Vasc. Biol.* 31: 842-850.

MONOS
Satisfaction
Guaranteed

Try **dystrophin (MANDRA1): sc-73592** or **dystrophin (H-5): sc-365954**, our highly recommended monoclonal alternatives to dystrophin (H-300). Also, for AC, HRP, FITC, PE, Alexa Fluor[®] 488 and Alexa Fluor[®] 647 conjugates, see **dystrophin (MANDRA1): sc-73592**.