

dystrophin (C-20): sc-7461

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 (C-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the C-terminus 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.

Blocking peptide available for competition studies, sc-7461 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

dystrophin (C-20) 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

dystrophin (C-20) is also recommended for detection of dystrophin in additional species, including canine, bovine, porcine and avian.

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: A-10 cell lysate: sc-3806, L8 cell lysate: sc-3807 or HeLa whole cell lysate: sc-2200.

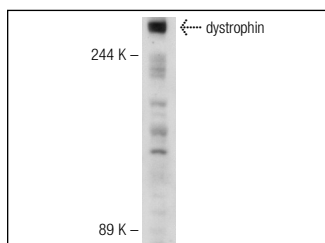
STORAGE

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

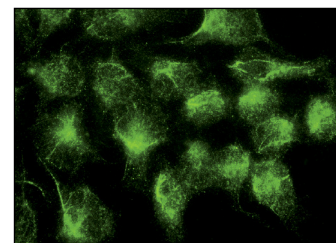
RESEARCH USE

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

DATA



dystrophin (C-20): sc-7461. Western blot analysis of dystrophin expression in HeLa whole cell lysate.



dystrophin (C-20): sc-7461: Immunofluorescence staining of methanol-fixed HeLa cells showing cytoskeletal localization.

SELECT PRODUCT CITATIONS

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- Xiong, D., et al. 2007. Inducible cardiac-restricted expression of enteroviral protease 2A is sufficient to induce dilated cardiomyopathy. *Circulation* 115: 94-102.
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- Yadava, R.S., et al. 2008. RNA toxicity in myotonic muscular dystrophy induces Nkx-2.5 expression. *Nat. Genet.* 40: 61-68.
- van Putten, M., et al. 2012. Comparison of skeletal muscle pathology and motor function of dystrophin and utrophin deficient mouse strains. *Neuromuscul. Disord.* 22: 406-417.
- van den Bergen, J.C., et al. 2013. Clinical characterisation of Becker muscular dystrophy patients predicts favourable outcome in exon-skipping therapy. *J. Neurol. Neurosurg. Psychiatr.* E-published.

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Try **dystrophin (MANDRA1): sc-73592** or **dystrophin (H-5): sc-365954**, our highly recommended monoclonal alternatives to dystrophin (C-20). Also, for AC, HRP, FITC, PE, Alexa Fluor® 488 and Alexa Fluor® 647 conjugates, see **dystrophin (MANDRA1): sc-73592**.