

dystrophin (4C7): sc-33697

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.

REFERENCES

1. Durbeej, M., et al. 2002. Muscular dystrophies involving the dystrophin-glycoprotein complex: an overview of current mouse models. *Curr. Opin. Genet. Dev.* 12: 349-361.
2. Michele, D.E., et al. 2003. Dystrophin-glycoprotein complex: post-translational processing and dystroglycan function. *J. Biol. Chem.* 278: 15457-15460.
3. Oak, S.A., et al. 2003. Skeletal muscle signaling pathway through the dystrophin glycoprotein complex and Rac1. *J. Biol. Chem.* 278: 39287-39295.
4. Johnson, B.D., et al. 2005. Convergent regulation of skeletal muscle Ca²⁺ channels by dystrophin, the actin cytoskeleton, and cAMP-dependent protein kinase. *Proc. Natl. Acad. Sci. USA* 102: 4191-4196.
5. Bhasin, N., et al. 2005. Molecular extensibility of mini-dystrophins and a dystrophin rod construct. *J. Mol. Biol.* 352: 795-806.

CHROMOSOMAL LOCATION

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

SOURCE

dystrophin (4C7) is a mouse monoclonal antibody raised against amino acids 1-68 of recombinant dystrophin of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

dystrophin (4C7) is available conjugated to agarose (sc-33697 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-33697 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-33697 PE), fluorescein (sc-33697 FITC), Alexa Fluor® 488 (sc-33697 AF488), Alexa Fluor® 546 (sc-33697 AF546), Alexa Fluor® 594 (sc-33697 AF594) or Alexa Fluor® 647 (sc-33697 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-33697 AF680) or Alexa Fluor® 790 (sc-33697 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

Alexa Fluor® is a trademark of Molecular Probes, Inc., Oregon, USA

APPLICATIONS

dystrophin (4C7) 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)] and immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

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.

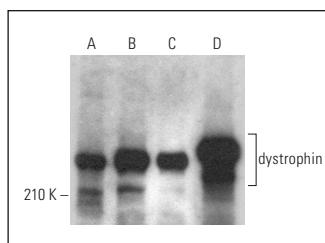
Molecular Weight of dystrophin: 427 kDa.

Positive Controls: C6 whole cell lysate: sc-364373, F9 cell lysate: sc-2245 or HeLa whole cell lysate: sc-2200.

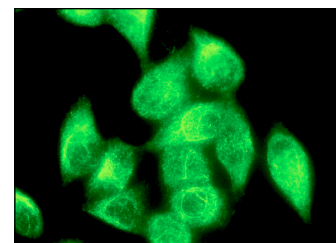
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



dystrophin (4C7): sc-33697. Western blot analysis of dystrophin expression in C6 (A), F9 (B), c4 (C) and HeLa (D) whole cell lysates.



dystrophin (4C7): sc-33697. Immunofluorescence staining of methanol-fixed HeLa cells showing cytoskeletal localization.

SELECT PRODUCT CITATIONS

1. Gonzalez-Hilarion, S., et al. 2012. Rescue of nonsense mutations by amlexanox in human cells. *Orphanet. J. Rare Dis.* 7: 56.

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.