SANTA CRUZ BIOTECHNOLOGY, INC.

dystrophin (4C7): sc-33697

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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 (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.

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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.

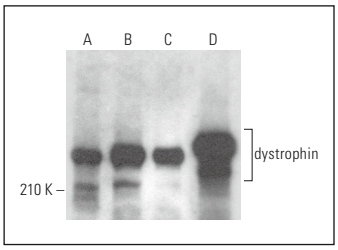
Molecular Weight of dystrophin: 427 kDa.

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

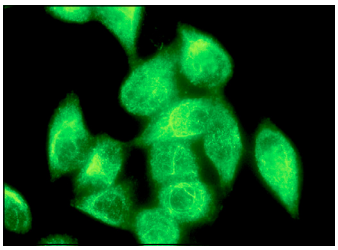
RECOMMENDED SUPPORT REAGENTS

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: 58.
2. Potter, R.A., et al. 2021. Dose-escalation study of systemically delivered rAAVrh74.MHCK7.micro-dystrophin in the mdx mouse model of Duchenne muscular dystrophy. Hum. Gene Ther. 32: 375-389.
3. Potter, R.A., et al. 2023. Expression and function of four AAV-based constructs for dystrophin restoration in the mdx mouse model of Duchenne muscular dystrophy. Biol. Open 12: bio059797.
4. Soussi, S., et al. 2023. IPSC derived cardiac fibroblasts of DMD patients show compromised actin microfilaments, metabolic shift and pro-fibrotic phenotype. Biol. Direct. 18: 41.

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.

PROTOCOLS

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