

dystrophin (MANDYS8): sc-58754

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.

CHROMOSOMAL LOCATION

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

SOURCE

dystrophin (MANDYS8) is a mouse monoclonal antibody raised against amino acids 816-1749 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.

APPLICATIONS

dystrophin (MANDYS8) 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 immunohistochemistry (including paraffin-embedded sections) (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.

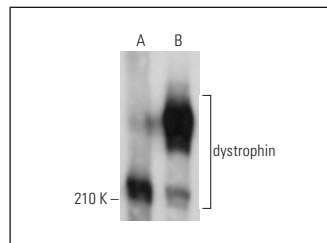
Molecular Weight of dystrophin: 427 kDa.

Positive Controls: human heart extract: sc-363763 or human fetal muscle tissue extract.

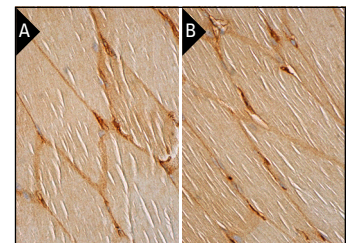
RECOMMENDED SUPPORT REAGENTS

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. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



dystrophin (MANDYS8): sc-58754. Western blot analysis of dystrophin expression in human heart (A) and human fetal muscle (B) tissue extracts.



dystrophin (MANDYS8): sc-58754. Immunoperoxidase staining of formalin fixed, paraffin-embedded mouse skeletal muscle (A) and rat skeletal muscle (B) tissue showing membrane and cytoplasmic staining of myocytes.

SELECT PRODUCT CITATIONS

1. Bishop, D.P., et al. 2021. Quantitative immuno-mass spectrometry imaging of skeletal muscle dystrophin. *Sci. Rep.* 11: 1128.
2. Mello, M.G., et al. 2021. Assessing the reproducibility of labelled antibody binding in quantitative multiplexed immuno-mass spectrometry imaging. *Anal. Bioanal. Chem.* 413: 5509-5516.

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.



See **dystrophin (MANDRA1): sc-73592** for dystrophin antibody conjugates, including AC, HRP, FITC, PE, and Alexa Fluor® 488, 546, 594, 647, 680 and 790.