

# 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<sub>2b</sub> 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.

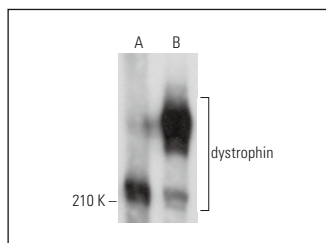
Molecular Weight of dystrophin: 427 kDa.

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

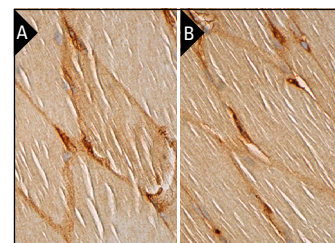
## RECOMMENDED SUPPORT REAGENTS

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. Westerhausen, M.T., et al. 2019. Super-resolution reconstruction for two- and three-dimensional LA-ICP-MS bioimaging. *Anal. Chem.* 91: 14879-14886.
2. Bishop, D.P., et al. 2021. Quantitative immuno-mass spectrometry imaging of skeletal muscle dystrophin. *Sci. Rep.* 11: 1128.
3. 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.



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