SANTA CRUZ BIOTECHNOLOGY, INC.

Myotilin (B-3): sc-393958

BACKGROUND

Myotilin, a sarcomeric protein that is encoded by the gene mapping to human chromosome 5q31.2, binds to α-actinin and is localized in the Z-line of myofibrils. Myotilin is expressed in skeletal and cardiac muscle, and it co-localizes with α-actinin in the sarcomeric I-bands where it directly interacts with α-actinin. Defects in the Myotilin gene are reported to cause a form of autosomal dominant limb-girdle muscular dystrophy (LGMD). Symptoms of adult onset LGMD are progressive weakness of the hip and shoulder girdles as well as a distinctive dysarthric pattern of speech. The muscle of affected individuals with LGMD shows degeneration of myofibers, variations in fiber size, fiber splitting, centrally located myonuclei and an enhanced number of autophagic vesicles.

REFERENCES

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3. Salmikangas, P., Mykkanen, O.M., Gronholm, M., Heiska, L., Kere, J. and Carpen, O. 1999. Myotilin, a novel sarcomeric protein with two Ig-like domains, is encoded by a candidate gene for limb-girdle muscular dystrophy. Hum. Mol. Genet. 8: 1329-1336.
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CHROMOSOMAL LOCATION

Genetic locus: MYOT (human) mapping to 5q31.2.

SOURCE

Myotilin (B-3) is a mouse monoclonal antibody raised against amino acids 331-416 mapping within an internal region of Myotilin of human origin.

PRODUCT

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

APPLICATIONS

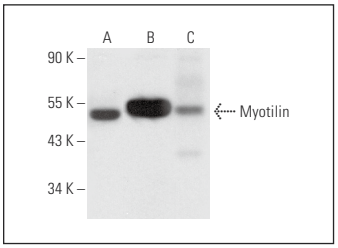
Myotilin (B-3) is recommended for detection of Myotilin of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for Myotilin siRNA (h): sc-43408, Myotilin shRNA Plasmid (h): sc-43408-SH and Myotilin shRNA (h) Lentiviral Particles: sc-43408-V.

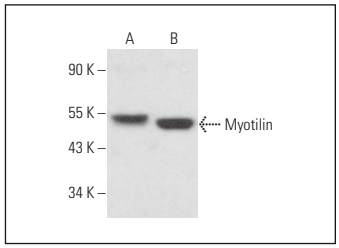
Molecular Weight of Myotilin: 57 kDa.

Positive Controls: A-673 cell lysate: sc-2414, human heart extract: sc-363763 or A549 cell lysate: sc-2413.

DATA



Myotilin (B-3): sc-393958. Western blot analysis of Myotilin expression in CCRF-CEM (**A**) and A549 (**B**) whole cell lysates and human heart tissue extract (**C**).



Myotilin (B-3): sc-393958. Western blot analysis of Myotilin expression in A549 (**A**) and A-673 (**B**) whole cell lysates.

SELECT PRODUCT CITATIONS

1. Sterzynska, K., Klejewski, A., Wojtowicz, K., Swierczewska, M., Nowicki, M., Brazert, J. and Januchowski, R. 2018. Myotilin, a new topotecan resistant protein in ovarian cancer cell lines. J. Cancer 9: 4413-4421.
2. Wojtowicz, K., Sterzynska, K., Swierczewska, M., Nowicki, M., Zabel, M. and Januchowski, R. 2021. Piperine targets different drug resistance mechanisms in human ovarian cancer cell lines leading to increased sensitivity to cytotoxic drugs. Int. J. Mol. Sci. 22: 4243.
3. Amrute-Nayak, M., Gand, L.V., Khan, B., Holler, T., Kefalakes, E., Kosanke, M., Kraft, T. and Nayak, A. 2022. SENP7 deSUMOylase-governed transcriptional program coordinates sarcomere assembly and is targeted in muscle atrophy. Cell Rep. 41: 111702.

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.