

Myotilin (E-10): sc-393957



The Power to Question

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

1. Speer, M.C., et al. 1995. Evidence for locus heterogeneity in autosomal dominant limb-girdle muscular dystrophy. *Am. J. Hum. Genet.* 57: 1371-1376.
2. Minetti, C., et al. 1998. Mutations in the caveolin-3 gene cause autosomal dominant limb-girdle muscular dystrophy. *Nat. Genet.* 18: 365-368.
3. Salmikangas, P., et al. 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.
4. Hauser, M.A., et al. 2000. Myotilin is mutated in limb girdle muscular dystrophy 1A. *Hum. Mol. Genet.* 9: 2141-2147.

CHROMOSOMAL LOCATION

Genetic locus: MYOT (human) mapping to 5q31.2; Myot (mouse) mapping to 18 B3.

SOURCE

Myotilin (E-10) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 321-420 within an internal region of Myotilin of human origin.

PRODUCT

Each vial contains 200 μ g IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Myotilin (E-10) is available conjugated to agarose (sc-393957 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-393957 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-393957 PE), fluorescein (sc-393957 FITC), Alexa Fluor[®] 488 (sc-393957 AF488), Alexa Fluor[®] 546 (sc-393957 AF546), Alexa Fluor[®] 594 (sc-393957 AF594) or Alexa Fluor[®] 647 (sc-393957 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor[®] 680 (sc-393957 AF680) or Alexa Fluor[®] 790 (sc-393957 AF790), 200 μ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

Blocking peptide available for competition studies, sc-393957 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

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APPLICATIONS

Myotilin (E-10) is recommended for detection of Myotilin of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g 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).

Myotilin (E-10) is also recommended for detection of Myotilin in additional species, including canine.

Suitable for use as control antibody for Myotilin siRNA (h): sc-43408, Myotilin siRNA (m): sc-43409, Myotilin shRNA Plasmid (h): sc-43408-SH, Myotilin shRNA Plasmid (m): sc-43409-SH, Myotilin shRNA (h) Lentiviral Particles: sc-43408-V and Myotilin shRNA (m) Lentiviral Particles: sc-43409-V.

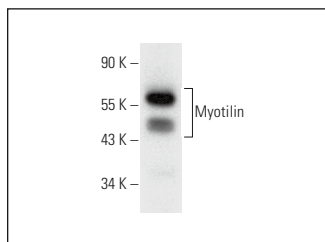
Molecular Weight of Myotilin: 57 kDa.

Positive Controls: rat skeletal muscle extract: sc-364810 or human skeletal muscle extract: sc-363776.

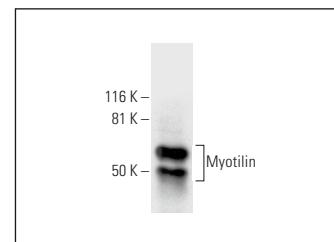
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ 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-IgG κ BP-FITC: sc-516140 or m-IgG κ 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



Myotilin (E-10): sc-393957. Western blot analysis of Myotilin expression in human skeletal muscle tissue extract.



Myotilin (E-10): sc-393957. Western blot analysis of Myotilin expression in rat skeletal muscle tissue extract.

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.