

myotubularin (T-16): sc-14782

BACKGROUND

X-linked recessive myotubular myopathy is a congenital muscular disease characterized by severe hypotonia and generalized muscle weakness that, in most cases, leads to early postnatal death. The gene responsible for myotubular myopathy MTM1 encodes a dual specificity phosphatase, named myotubularin, which is highly conserved through evolution. The gene for MTM1 is localized to a 300 kb critical region on human Xq28 between IDS and GRBRA3. Human MTM1, a 603 amino-acid protein, is mutated in myotubular myopathy. The largely related protein hMTMR2 is found mutated in a recessive form of Charcot-Marie-Tooth neuropathy. Myotubularin is primarily a lipid phosphatase that acts on phosphatidylinositol 3-monophosphate and is involved in the regulation of the phosphatidylinositol 3-kinase (PI 3-kinase) pathway and membrane trafficking. Wildtype myotubularin can directly dephosphorylate PI 3-P and PI 4-P *in vitro*. Thus, it decreases PI 3-P levels by down-regulating PI 3-K activity and by facilitating the degradation of PI 3-P.

CHROMOSOMAL LOCATION

Genetic locus: MTM1 (human) mapping to Xq28; Mtm1 (mouse) mapping to X A7.2.

SOURCE

myotubularin (T-16) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of myotubularin of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-14782 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

myotubularin (T-16) is recommended for detection of myotubularin of human and, to a lesser extent, mouse and rat 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 solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

myotubularin (T-16) is also recommended for detection of myotubularin in additional species, including canine and porcine.

Suitable for use as control antibody for myotubularin siRNA (h): sc-44356, myotubularin siRNA (m): sc-44357, myotubularin shRNA Plasmid (h): sc-44356-SH, myotubularin shRNA Plasmid (m): sc-44357-SH, myotubularin shRNA (h) Lentiviral Particles: sc-44356-V and myotubularin shRNA (m) Lentiviral Particles: sc-44357-V.

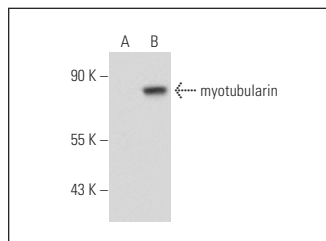
Molecular Weight of myotubularin: 66 kDa.

Positive Controls: myotubularin (h): 293 Lysate: sc-158751 or Hep G2 cell lysate: sc-2227.

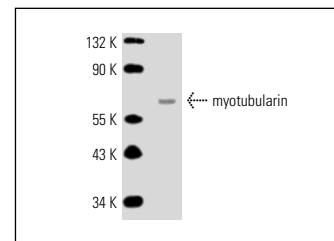
RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



myotubularin (T-16): sc-14782. Western blot analysis of myotubularin expression in non-transfected: sc-110760 (A) and human myotubularin transfected: sc-158751 (B) 293 whole cell lysates.



myotubularin (T-16): sc-14782. Western blot analysis of myotubularin expression in rat testis 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.

PROTOCOLS

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



Try **myotubularin (F-1): sc-377309**, our highly recommended monoclonal alternative to myotubularin (T-16).