SANTA CRUZ BIOTECHNOLOGY, INC.

Telethonin (G-11): sc-25327

SANTA CRUZ BIOTECHNOLOGY
The Power to Question

BACKGROUND

Titin, also known as connectin, is a large protein involved in the temporal and spatial control of the assembly of the highly ordered sarcomeres (contractile units) of striated muscle. In addition to sarcomere assembly Titin also functions to maintain the structural integrity of the contracting myofibrils within the muscle as well as organizing the machinery for condensation of chromosomes in dividing cells. Titin is a giant protein composed of 27,000 amino acids and contains an autoregulated serine kinase catalytic domain as well as a calcium/calmodulin binding region that are involved in its activation. Activated Titin phosphorylates the muscle protein Telethonin, a sarcomeric protein abundant in heart and skeletal muscle, implicating Titin activity in the reorganization of the cytoskeleton during myofibrillogenesis.

CHROMOSOMAL LOCATION

Genetic locus: TCAP (human) mapping to 17q12; Tcap (mouse) mapping to 11 D.

SOURCE

Telethonin (G-11) is a mouse monoclonal antibody raised against amino acids 58-167 of telethonin of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Telethonin (G-11) is available conjugated to agarose (sc-25327 AC), 500 µg/ 0.25 ml agarose in 1 ml, for IP; to HRP (sc-25327 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-25327 PE), fluorescein (sc-25327 FITC), Alexa Fluor® 488 (sc-25327 AF488), Alexa Fluor® 546 (sc-25327 AF546), Alexa Fluor® 594 (sc-25327 AF594) or Alexa Fluor® 647 (sc-25327 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-25327 AF680) or Alexa Fluor® 790 (sc-25327 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

Telethonin (G-11) is recommended for detection of Telethonin of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1,000), 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), immunohistochemistry (including paraffin-embedded sections) (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 Telethonin siRNA (h): sc-36638, Telethonin siRNA (m): sc-36637, Telethonin shRNA Plasmid (h): sc-36638-SH, Telethonin shRNA Plasmid (m): sc-36637-SH, Telethonin shRNA (h) Lentiviral Particles: sc-36638-V and Telethonin shRNA (m) Lentiviral Particles: sc-36637-V.

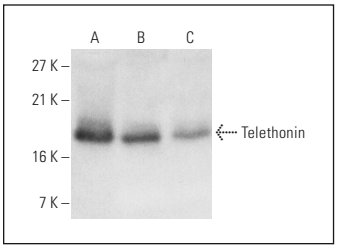
Molecular Weight of Telethonin: 19 kDa.

Positive Controls: mouse heart extract: sc-2254, rat heart extract: sc-2393 or rat skeletal muscle extract: sc-364810.

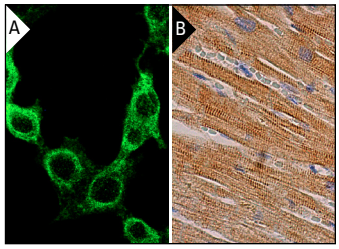
STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

DATA



Telethonin (G-11): sc-25327. Western blot analysis of Telethonin expression in rat skeletal muscle (**A**), mouse heart (**B**) and rat heart (**C**) tissue extracts.



Telethonin (G-11): sc-25327. Immunofluorescence staining of methanol-fixed Sol8 cells showing cytoplasmic localization (**A**). Immunoperoxidase staining of formalin fixed, paraffin-embedded human heart muscle tissue showing cytoplasmic staining of myocytes (**B**).

SELECT PRODUCT CITATIONS

1. Heng, A.E., et al. 2008. Coordinate expression of the 19S regulatory complex and evidence for ubiquitin-dependent Telethonin degradation in the unloaded soleus muscle. Int. J. Biochem. Cell Biol. 40: 2544-2552.
2. Claeys, K.G., et al. 2010. Myopathy with hexagonally cross-linked crystalloid inclusions: delineation of a clinico-pathological entity. Neuromuscul. Disord. 20: 701-708.
3. Ferreiro, A., et al. 2011. Telethonin-deficiency initially presenting as a congenital muscular dystrophy. Neuromuscul. Disord. 21: 433-438.
4. Paim, J.F., et al. 2013. Muscle phenotypic variability in limb girdle muscular dystrophy 2G. J. Mol. Neurosci. 50: 339-344.
5. Cotta, A., et al. 2014. Common recessive limb girdle muscular dystrophies differential diagnosis: why and how? Arq. Neuropsiquiatr. 72: 721-734.
6. Barresi, R., et al. 2015. Conserved expression of truncated Telethonin in a patient with limb-girdle muscular dystrophy 2G. Neuromuscul. Disord. 25: 349-352.
7. de Fuenmayor-Fernández de la Hoz, C.P., et al. 2016. Novel mutation in TCAP manifesting with asymmetric calves and early-onset joint retractions. Neuromuscul. Disord. 26: 749-753.
8. Zuppinger, C., et al. 2017. Characterization of cytoskeleton features and maturation status of cultured human iPSC-derived cardiomyocytes. Eur. J. Histochem. 61: 2763.
9. Iyer, S., et al. 2019. Precise therapeutic gene correction by a simple nuclease-induced double-stranded break. Nature 568: 561-565.
10. Lv, X., et al. 2021. Distal myopathy due to TCAP variants in four unrelated Chinese patients. Neurogenetics 22: 1-10.

RESEARCH USE

For research use only, not for use in diagnostic procedures.