SANTA CRUZ BIOTECHNOLOGY, INC.

α-dystroglycan (IIH6): sc-53987

BACKGROUND

Dystroglycan (DG) is a cell surface receptor for several extracellular matrix molecules including laminins, Agrin and Perlecan. Dystroglycan function is required for the formation of basement membranes in early development and the organization of Laminin on the cell surface. α-dystroglycan is a membrane- associated, extracellular glycoprotein that is anchored to the cell-membrane by binding to the transmembrane glycoprotein β-dystroglycan to form an α/β-dystroglycan-complex. Additionally, dystroglycan is part of a multimolecular complex, where it associates with dystrophin, at the sarcolemma, to form the dystrophin-associated protein complex, or with utrophin, at the neuromuscular junction, to form the utrophin-associated protein complex. Dystroglycan is also thought to participate in the clustering of nicotinic acetylcholine receptors at the neuromuscular junction.

CHROMOSOMAL LOCATION

Genetic locus: DAG1 (human) mapping to 3p21.31; Dag1 (mouse) mapping to 9 F2.

SOURCE

α-dystroglycan (IIH6) is a mouse monoclonal antibody raised against purified dystrophin-glycoprotein complex of rabbit origin.

PRODUCT

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

APPLICATIONS

α-dystroglycan (IIH6) is recommended for detection of α-dystroglycan of mouse, rat, human, rabbit and canine 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 α/β-dystroglycan siRNA (h): sc-43488, α/β-dystroglycan siRNA (m): sc-43489, α/β-dystroglycan shRNA Plasmid (h): sc-43488-SH, α/β-dystroglycan shRNA Plasmid (m): sc-43489-SH, α/β-dystroglycan shRNA (h) Lentiviral Particles: sc-43488-V and α/β-dystroglycan shRNA (m) Lentiviral Particles: sc-43489-V.

Molecular Weight of α-dystroglycan skeletal muscle: 156 kDa.

Molecular Weight of α-dystroglycan brain: 120 kDa.

Positive Controls: rat skeletal muscle extract: sc-364810, mouse skeletal muscle extract: sc-364250 or human skeletal muscle extract: sc-363776.

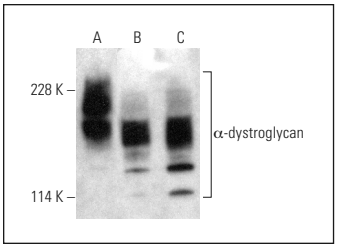
STORAGE

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

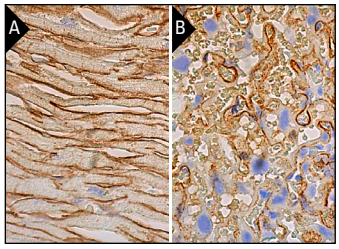
RESEARCH USE

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

DATA



α-dystroglycan (IIH6) HRP: sc-53987 HRP. Direct western blot analysis of α-dystroglycan expression in human skeletal muscle (**A**), mouse skeletal muscle (**B**) and rat skeletal muscle (**C**) tissue extracts.



α-dystroglycan (IIH6): sc-53987. Immunoperoxidase staining of formalin fixed, paraffin-embedded human heart muscle tissue showing membrane staining of myocytes (**A**). Immunoperoxidase staining of formalin fixed, paraffin-embedded mouse placenta tissue showing membrane staining of trophoblastic cells (**B**).

SELECT PRODUCT CITATIONS

1. Williams, S. and Jacobson, C. 2010. α-dystroglycan is essential for the induction of Egr3, a transcription factor important in muscle spindle formation. Dev. Neurobiol. 70: 498-507.
2. Munoz, J., et al. 2010. LG4-5 domains of laminin-211 binds α-dystroglycan to allow myotube attachment and prevent anoikis. J. Cell. Physiol. 222: 111-119.
3. Huang, Q., et al. 2015. The glycosyltransferase LARGE2 is repressed by Snail and ZEB1 in prostate cancer. Cancer Biol. Ther. 16: 125-136.
4. Parvatiyar, M.S., et al. 2015. Sarcospan regulates cardiac isoproterenol response and prevents duchenne muscular dystrophy-associated cardiomyopathy. J. Am. Heart Assoc. 23: e002481.
5. Marshall, J.L., et al. 2015. Sarcospan integration into laminin-binding adhesion complexes that ameliorate muscular dystrophy requires utrophin and α7 integrin. Hum. Mol. Genet. 24: 2011-2022.
6. Eldridge, S., et al. 2016. Agrin mediates chondrocyte homeostasis and requires both LRP4 and α-dystroglycan to enhance cartilage formation *in vitro* and *in vivo*. Ann. Rheum. Dis. 75: 1228-1235.
7. Gibbs, E.M., et al. 2016. High levels of sarcospan are well tolerated and act as a sarcolemmal stabilizer to address skeletal muscle and pulmonary dysfunction in DMD. Hum. Mol. Genet. 25: 5395-5406.
8. Bauché, S., et al. 2017. Mutations in GFPT1-related congenital myasthenic syndromes are associated with synaptic morphological defects and underlie a tubular aggregate myopathy with synaptopathy. J. Neurol. 264: 1791-1803.

PROTOCOLS

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