α -dystroglycan (2238E2D2): sc-65629



The Power to Question

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 acetyl-choline receptors at the neuromuscular junction.

REFERENCES

- Cote, P.D., Moukhles, H., Lindenbaum, M. and Carbonetto, S. 1999. Chimaeric mice deficient in dystroglycans develop muscular dystrophy and have disrupted myoneural synapses. Nat. Genet. 23: 338-342.
- Seifert, J., Ogawa, T., Kurono, S. and Ito, Y. 2000. Syntheses of α-dystroglycan derived glycosyl amino acids carrying a novel mannosyl serine/threonine linkage. Glycoconi. J. 17: 407-423.
- 3. Henry, M.D., Satz, J.S., Brakebusch, C., Costell, M., Gustafsson, E., Fassler, R. and Campbell, K.P. 2001. Distinct roles for dystroglycan, β1 Integrin and Perlecan in cell surface Laminin organization. J. Cell Sci. 114: 1137-1144.
- Masaki, T., Matsumura, K., Hirata, A., Yamada, H., Hase, A., Shimizu, T., Yorifuji, H., Motoyoshi, K. and Kamakura, K. 2001. Expression of dystroglycan complex in satellite cells of dorsal root ganglia. Acta Neuropathol. 101: 174-178.
- Marchand, S., Stetzkowski-Marden, F. and Cartaud, J. 2001. Differential targeting of components of the dystrophin complex to the postsynaptic membrane. Eur. J. Neurosci. 13: 221-229.
- Bonuccelli, G., Sotgia, F., Capozza, F., Gazzerro, E., Minetti, C. and Lisanti, M.P. 2007. Localized treatment with a novel FDA-approved proteasome inhibitor blocks the degradation of dystrophin and dystrophin-associated proteins in mdx mice. Cell Cycle 6: 1242-1248.
- Prados, B., Pena, A., Cotarelo, R.P., Valero, M.C. and Cruces, J. 2007. Expression of the murine Pomt1 gene in both the developing brain and adult muscle tissues and its relationship with clinical aspects of Walker-Warburg syndrome. Am. J. Pathol. 170: 1659-1668.
- 8. Darin, N., Kroksmark, A.K., Ahlander, A.C., Moslemi, A.R., Oldfors, A. and Tulinius, M. 2007. Inflammation and response to steroid treatment in limb-girdle muscular dystrophy 2l. Eur. J. Paediatr. Neurol. 6: 353-357
- Sciandra, F., Gawlik, K.I., Brancaccio, A. and Durbeej, M. 2007.
 Dystroglycan: a possible mediator for reducing congenital muscular dystrophy? Trends Biotechnol. 25: 262-268.

CHROMOSOMAL LOCATION

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

SOURCE

 α -dystroglycan (2237E2D2) is a mouse monoclonal antibody raised against brain tissue homogenate, with epitope mapping to the glycosylation site of α -dystroglycan of bovine origin.

PRODUCT

Each vial contains 200 μ g IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

 $\alpha\text{-dystroglycan}$ (2238E2D2) is recommended for detection of brain $\alpha\text{-dystroglycan}$ of mouse, rat, human, bovine and rabbit origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000); non cross-reactive with skeletal muscle $\alpha\text{-dystroglycan}.$

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 skeletal muscle α -dystroglycan: 156 kDa.

Molecular Weight of brain α -dystroglycan: 120 kDa. Positive Controls: mouse brain tissue extract: sc-2253.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-lgG κ BP-HRP: sc-516102 or m-lgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz MarkerTM Molecular Weight Standards: sc-2035, UltraCruz[®] Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048.

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 for detailed protocols and support products.



See α -dystroglycan (IIH6): sc-53987 for α -dystroglycan antibody conjugates, including AC, HRP, FITC, PE, and Alexa Fluor[®] 488, 546, 594, 647, 680 and 790.

Santa Cruz Biotechnology, Inc. 1.800.457.3801 831.457.3800 fax 831.457.3801 **Europe** +00800 4573 8000 49 6221 4503 0 **www.scbt.com**