LARGE (C-17): sc-46342



The Power to Question

BACKGROUND

Glycosyltransferase-like protein LARGE, also designated acetylglucosaminyltransferase-like protein, belongs to the glycosyltransferase 8 family. This ubiquitously expressed protein is a type II membrane protein. Although it is widely expressed, highest levels of detection are in heart, brain and skeletal muscle. LARGE carries out the synthesis of glycosphingolipid and glycoprotein sugar chains and is part of the repeated disaccharide unit addition. It may also be important in the hyperglycosylation of α -dystroglycan. This interaction of LARGE with dystroglycan is crucial for the biosynthetic pathway to create functional dystroglycan. Loss of functional dystroglycan can result in muscle degeneration. The gene encoding for LARGE maps to chromosome 22q12.3, and defects in this gene can cause congenital muscular dystrophy, an autosomal recessive disorder. LARGE co-localizes with GM130, a Golgi marker.

REFERENCES

- 1. Grewal, P.K., et al. 2001. Mutant glycosyltransferase and altered glycosylation of α -dystroglycan in the myodystrophy mouse. Nat. Genet. 28: 151-154.
- Holzfeind, P.J., et al. 2002. Skeletal, cardiac and tongue muscle pathology, defective retinal transmission, and neuronal migration defects in the LARGE (myd) mouse defines a natural model for glycosylation-deficient muscleeye-brain disorders. Hum. Mol. Genet. 11: 2673-2687.
- Barresi, R., et al. 2004. LARGE can functionally bypass α-dystroglycan glycosylation defects in distinct congenital muscular dystrophies. Nat. Med. 10: 696-703.
- 4. Kanagawa, M., et al. 2004. Molecular recognition by LARGE is essential for expression of functional dystroglycan. Cell 117: 953-964.
- Brockington, M., et al. 2005. Localization and functional analysis of the LARGE family of glycosyltransferases: significance for muscular dystrophy. Hum. Mol. Genet. 14: 657-665.

CHROMOSOMAL LOCATION

Genetic locus: LARGE (human) mapping to 22q12.3; Large (mouse) mapping to 8 B3.3.

SOURCE

LARGE (C-17) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of LARGE of human origin.

PRODUCT

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

Blocking peptide available for competition studies, sc-46342 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

LARGE (C-17) is recommended for detection of LARGE of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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).

LARGE (C-17) is also recommended for detection of LARGE in additional species, including equine, canine, bovine, porcine and avian.

Suitable for use as control antibody for LARGE siRNA (h): sc-44965, LARGE siRNA (m): sc-44966, LARGE shRNA Plasmid (h): sc-44965-SH, LARGE shRNA Plasmid (m): sc-44966-SH, LARGE shRNA (h) Lentiviral Particles: sc-44965-V and LARGE shRNA (m) Lentiviral Particles: sc-44966-V.

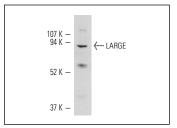
Molecular Weight of LARGE: 88 kDa.

Positive Controls: NIH/3T3 whole cell lysate: sc-2210 or C2C12 whole cell lysate: sc-364188.

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



LARGE (C-17): sc-46342. Western blot analysis of LARGE expression in NIH/3T3 whole cell lysate.

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.

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