SANTA CRUZ BIOTECHNOLOGY, INC.

LARGE (H-164): sc-99164

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.
2. 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 muscle-eye-brain disorders. Hum. Mol. Genet. 11: 2673-2687.
3. 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.
5. 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, GYLTL1B (human) mapping to11p11.2; Large (mouse) mapping to 8 B3.3, Gyltl1b (mouse) mapping to 2 E1.

SOURCE

LARGE (H-164) is a rabbit polyclonal antibody raised against amino acids 137-300 mapping within an internal region of LARGE 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.

STORAGE

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

PROTOCOLS

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

APPLICATIONS

LARGE (H-164) is recommended for detection of LARGE, and to a lesser extent LARGE2 of mouse, rat and human 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).

LARGE (H-164) is also recommended for detection of LARGE, and to a lesser extent, LARGE2 in additional species, including equine, canine, bovine and avian.

Molecular Weight of LARGE: 88 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

RESEARCH USE

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