SANTA CRUZ BIOTECHNOLOGY, INC.

POMGnT1 (N-20): sc-23356

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

The product of the POMGNT1 gene, protein O-mannose β-1, 2-N-acetylglucosaminyltransferase, participates in O-mannosyl glycan synthesis. POMGnT1 is an N(in)/C(out) (type II) membrane protein localized in the medial-Golgi that initiates the conversion of high mannose N-glycans to complex N-glycans. Specifically, POMGnT1 is a glycosylation enzyme that participates in the synthesis of O-mannosyl glycan, a laminin-binding ligand of α-dystroglycan that is rarely synthesized in mammals. Mutations in the POMGNT1 gene cause muscle-eye-brain disease (MEB), an autosomal recessive disorder characterized by congenital muscular dystrophy, ocular abnormalities and lissencephaly. Altered glycosylation of α-dystroglycan may play a critical role in the pathomechanism of MEB as well as Walker-Warburg syndrome (WWS), characterized by the absence of glycosylation of α-dystroglycan. The human POMGnT1 gene maps to chromosome 1p34.1 and encodes a 660-amino acid type II transmembrane protein.

REFERENCES

1. Burke, J., et al. 1992. The transmembrane and flanking sequences of β-1, 2-N-acetylglucosaminyltransferase I specify medial-Golgi localization. J. Biol. Chem. 267: 24433-24440.
2. Yoshida, A., et al. 2001. Muscular dystrophy and neuronal migration disorder caused by mutations in a glycosyltransferase, POMGnT1. Dev. Cell 1: 717-724.
3. Kano, H., et al. 2002. Deficiency of α-dystroglycan in muscle-eye-brain disease. Biochem. Biophys. Res. Commun. 291: 1283-1286.
4. Michele, D.E., et al. 2002. Post-translational disruption of dystroglycan-ligand interactions in congenital muscular dystrophies. Nature 418: 417-422.
5. Beltran-Valero De Bernabe, D., et al. 2002. Mutations in the O-mannosyltransferase gene POMT1 give rise to the severe neuronal migration disorder Walker-Warburg syndrome. Am. J. Hum. Genet. 71: 1033-1043.
6. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 606822. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/

CHROMOSOMAL LOCATION

Genetic locus: POMGNT1 (human) mapping to 1p34.1; Pomgnt1 (mouse) mapping to 4 D1.

SOURCE

POMGnT1 (N-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of POMGnT1 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.

Blocking peptide available for competition studies, sc-23356 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

POMGnT1 (N-20) is recommended for detection of POMGnT1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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).

POMGnT1 (N-20) is also recommended for detection of POMGnT1 in additional species, including canine, bovine, porcine and avian.

Suitable for use as control antibody for POMGnT1 siRNA (h): sc-40865, POMGnT1 siRNA (m): sc-40866, POMGnT1 shRNA Plasmid (h): sc-40865-SH, POMGnT1 shRNA Plasmid (m): sc-40866-SH, POMGnT1 shRNA (h) Lentiviral Particles: sc-40865-V and POMGnT1 shRNA (m) Lentiviral Particles: sc-40866-V.

Molecular Weight of POMGnT1: 75 kDa.

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

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) 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.

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 or our catalog for detailed protocols and support products.