SANTA CRUZ BIOTECHNOLOGY, INC.

Glucosidase I (H-300): sc-134706

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The Power to Question

BACKGROUND

Glycosylation of asparagine residues in Asn-X-Ser/Thr motifs in proteins commonly occur in the lumen of the endoplasmic reticulum (ER). Glucosidase I catalyzes the first step in the N-linked oligosaccharide processing pathway. It specifically removes the distal α 1,2-linked glucose residue from the Glc3-Man9-GlcNAc2 oligosaccharide precursor. Glucosidase I contains a short cytosolic tail, a single pass transmembrane domain and a large C-terminal catalytic domain located on the luminal side of the ER. Mutations in the gene encoding Glucosidase I result in the congenital disorder glycosylation (CDG-IIb), which is characterized by generalized hypotonia, dysmorphic features, hepatomegaly, hypoventilation, feeding problems, seizures and death. Two point mutations in the Glucosidase I gene have been identified and result in amino acid substitutions, namely Arg486Thr and Phe652Leu, that affect polypeptide folding and active site formation.

REFERENCES

1. Kalz-Füller, B., et al. 1995. Cloning and expression of Glucosidase I from human hippocampus. Eur. J. Biochem. 231: 344-351.
2. Khan, F.A., et al. 1999. Genomic organization and promoter activity of Glucosidase I gene. Glycobiology 9: 797-806.
3. De Praeter CM, G.J., et al. 2000. A novel disorder caused by defective biosynthesis of N-linked oligosaccharides due to Glucosidase I deficiency. Am. J. Hum. Genet. 66: 1744-1756.
4. Völker, C., et al. 2002. Processing of N-linked carbohydrate chains in a patient with Glucosidase I deficiency (CDG type IIb). Glycobiology 12: 473-483.
5. Hardt, B., et al. 2003. (Arg)3 within the N-terminal domain of Glucosidase I contains ER targeting information but is not required absolutely for ER localization. Glycobiology 13: 159-168.
6. Hong, Y., et al. 2004. The Lec23 Chinese hamster ovary mutant is a sensitive host for detecting mutations in α-Glucosidase I that give rise to congenital disorder of glycosylation IIb (CDG IIb). J. Biol. Chem. 279: 49894-49901.
7. Ruddock, L.W., et al. 2006. N-glycan processing in ER quality control. J. Cell Sci. 119: 4373-4380.

CHROMOSOMAL LOCATION

Genetic locus: MOGS (human) mapping to 2p13.1; Mogs (mouse) mapping to 6 C3.

SOURCE

Glucosidase I (H-300) is a rabbit polyclonal antibody raised against amino acids 395-694 mapping within an internal region of Glucosidase I 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.

APPLICATIONS

Glucosidase I (H-300) is recommended for detection of Glucosidase I 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).

Glucosidase I (H-300) is also recommended for detection of Glucosidase I in additional species, including equine, canine and porcine.

Suitable for use as control antibody for Glucosidase I siRNA (h): sc-94835, Glucosidase I siRNA (m): sc-145445, Glucosidase I shRNA Plasmid (h): sc-94835-SH, Glucosidase I shRNA Plasmid (m): sc-145445-SH, Glucosidase I shRNA (h) Lentiviral Particles: sc-94835-V and Glucosidase I shRNA (m) Lentiviral Particles: sc-145445-V.

Molecular Weight of Glucosidase I: 92 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.

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.