SANTA CRUZ BIOTECHNOLOGY, INC.

Glucosidase I (H-6): sc-365399

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, C.M., 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. and Molinari, M. 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-6) is a mouse monoclonal 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_1 kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

Glucosidase I (H-6) is recommended for detection of Glucosidase I of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

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.

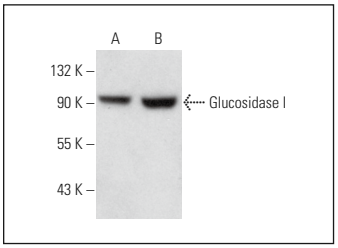
Molecular Weight of Glucosidase I: 92 kDa.

Positive Controls: H4 cell lysate: sc-2408 or COLO 320DM cell lysate: sc-2226.

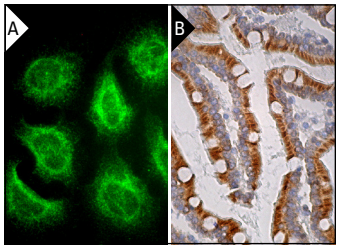
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 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 m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



Glucosidase I (H-6): sc-365399. Western blot analysis of Glucosidase I expression in H4 (**A**) and COLO 320DM (**B**) whole cell lysates.



Glucosidase I (H-6): sc-365399. Immunofluorescence staining of methanol-fixed HeLa cells showing cytoplasmic localization (**A**). Immunoperoxidase staining of formalin fixed, paraffin-embedded human duodenum tissue showing cytoplasmic staining of glandular cells (**B**).

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.