

Glucosidase I (N-20): sc-130146

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 Arg 486-Thr and Phe 652-Leu, that affect polypeptide folding and active site formation.

REFERENCES

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CHROMOSOMAL LOCATION

Genetic locus: GCS1 (human) mapping to 2p13.1.

SOURCE

Glucosidase I (N-20) is a purified rabbit polyclonal antibody raised against a peptide mapping near the N-terminus of Glucosidase I of human origin.

RESEARCH USE

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

PRODUCT

Each vial contains 100 μ g IgG in 1.0 ml PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

Glucosidase I (N-20) is recommended for detection of Glucosidase I of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

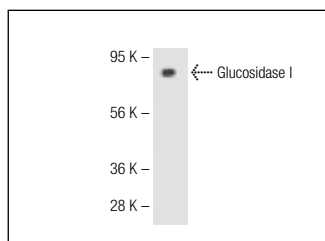
Suitable for use as control antibody for Glucosidase I siRNA (h): sc-94835, Glucosidase I shRNA Plasmid (h): sc-94835-SH and Glucosidase I shRNA (h) Lentiviral Particles: sc-94835-V.

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.

DATA



Glucosidase I (N-20): sc-130146. Western blot analysis of Glucosidase I expression in 293 whole cell lysate.

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.

MONOS
Satisfaction
Guaranteed

Try **Glucosidase I (C-11): sc-374006** or **Glucosidase I (H-6): sc-365399**, our highly recommended monoclonal alternatives to Glucosidase I (N-20).