**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 $\alpha$-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 Arg486 Thr and Phe652 Leu, that affect polypeptide folding and active site formation.

**REFERENCES**


**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 IgG1 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: HeLa whole cell lysate: sc-2200, 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-IgG $\kappa$BP-HRP: sc-516102 or m-IgG $\kappa$BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), 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 m-IgG $\kappa$BP-FITC: sc-516140 or m-IgG $\kappa$BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

**DATA**

**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.