SANTA CRUZ BIOTECHNOLOGY, INC.

GNPTAB (S-13): sc-107566

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

GlcNAc-1-phosphotransferase subunits α/β (GNPTAB), also known as N-acetylglucosamine-1-phosphotransferase subunits α/β or UDP-N-acetylglucosamine-1-phosphotransferase subunits α/β, is a 1,256 amino acid member of the stealth family of proteins. Localized to the Golgi apparatus membrane, GNPTAB is expressed in heart, brain, placenta, lung, liver, kidney, pancreas and skeletal muscle. GNPTAB catalyzes the formation of mannose 6-phosphate (M6P) markers on high mannose type oligosaccharides in the Golgi apparatus. M6Ps bind to the M6P receptors (MPR), after which MPRs can mediate the vesicular transport of lysosomal enzymes to the endosomal/prelysosomal compartment. Defects in the gene encoding GNPTAB lead to mucolipidosis type II (MLII), also known as inclusion cell disease (ICD), and mucolipidosis type III complementation group A (MLIIIA), also known as variant pseudo-Hurler polydystrophy. Two isoforms of GNPTAB exist as a result of alternative splicing events.

REFERENCES

1. Dittmer, F., et al. 1999. Phosphorylation of arylsulphatase A occurs through multiple interactions with the UDP-N-acetylglucosamine-1-phosphotransferase proximal and distal to its retrieval site by the KDEL receptor. Biochem. J. 340: 729-736.
2. Tiede, S., et al. 2004. A novel mutation in UDP-N-acetylglucosamine-1-phosphotransferase gamma subunit (GNPTAG) in two siblings with mucolipidosis type III alters a used glycosylation site. Hum. Mutat. 24: 535.
3. Raas-Rothschild, A., et al. 2004. Genomic organisation of the UDP-N-acetylglucosamine-1-phosphotransferase gamma subunit (GNPTAG) and its mutations in mucolipidosis III. J. Med. Genet. 41: e52.
4. Tiede, S., et al. 2005. Missense mutations in N-acetylglucosamine-1-phosphotransferase α/β subunit gene in a patient with mucolipidosis III and a mild clinical phenotype. Am. J. Med. Genet. A 137A: 235-240.
5. Tiede, S., et al. 2005. Mucolipidosis II is caused by mutations in GNPTA encoding the α/β GlcNAc-1-phosphotransferase. Nat. Med. 11: 1109-1112.

CHROMOSOMAL LOCATION

Genetic locus: GNPTAB (human) mapping to 12q23.2; Gnptab (mouse) mapping to 10 C1.

SOURCE

GNPTAB (S-13) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of GNPTAB 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-107566 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

RESEARCH USE

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

APPLICATIONS

GNPTAB (S-13) is recommended for detection of GNPTAB 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).

GNPTAB (S-13) is also recommended for detection of GNPTAB in additional species, including equine, canine, bovine, porcine and avian.

Suitable for use as control antibody for GNPTAB siRNA (h): sc-95992, GNPTAB siRNA (m): sc-145658, GNPTAB shRNA Plasmid (h): sc-95992-SH, GNPTAB shRNA Plasmid (m): sc-145658-SH, GNPTAB shRNA (h) Lentiviral Particles: sc-95992-V and GNPTAB shRNA (m) Lentiviral Particles: sc-145658-V.

Molecular Weight of GNPTAB: 144 kDa.

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.

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.