SANTA CRUZ BIOTECHNOLOGY, INC.

ALG6 (Z164): sc-100506

BACKGROUND

ALG6 (asparagine-linked glycosylation 6 homolog), is a 507 amino acid member of the ALG6/ALG8 glucosyltransferase family that functions as an α1,3-glucosyltransferase required for proper asparagine-linked glycosylation of proteins. ALG6 is a multi-pass membrane protein that localizes to the endoplasmic reticulum (ER). Specifically, ALG6 adds the first of three glucose residues added to dolichylpyrophosphate-linked oligosaccharide, a precursor for N-linked glycosylation. Mutations in the gene encoding ALG6 disrupt protein glycosylation and result in congenital disorder of glycosylation type 1C (CDG1C). CDG1C is a multisystem disease characterized by under-glycosylated serum proteins. Patients with CDG1C exhibit delayed statomotor development, are mentally retarded and have muscular hypotonia.

REFERENCES

1. Imbach, T., Burda, P., Kuhnert, P., Wevers, R.A., Aebi, M., Berger, E.G. and Hennet, T. 1999. A mutation in the human ortholog of the *Saccharomyces cerevisiae* ALG6 gene causes carbohydrate-deficient glycoprotein syndrome type-1C. Proc. Natl. Acad. Sci. USA 96: 6982-6987.
2. Westphal, V., Murch, S., Kim, S., Srikrishna, G., Winchester, B., Day, R. and Freeze, H.H. 2000. Reduced heparan sulfate accumulation in enterocytes contributes to protein-losing enteropathy in a congenital disorder of glycosylation. Am. J. Pathol. 157: 1917-1925.
3. Westphal, V., Schottstädt, C., Marquardt, T. and Freeze, H.H. 2000. Analysis of multiple mutations in the hALG6 gene in a patient with congenital disorder of glycosylation 1C. Mol. Genet. Metab. 70: 219-223.
4. Freeze, H.H. and Westphal, V. 2001. Balancing N-linked glycosylation to avoid disease. Biochimie 83: 791-799.
5. Oriol, R., Martinez-Duncker, I., Chantret, I., Mollicone, R. and Codogno, P. 2002. Common origin and evolution of glycosyltransferases using Dol-P-monosaccharides as donor substrate. Mol. Biol. Evol. 19: 1451-1463.
6. Westphal, V., Xiao, M., Kwok, P.Y. and Freeze, H.H. 2003. Identification of a frequent variant in ALG6, the cause of congenital disorder of glycosylation-1C. Hum. Mutat. 22: 420-421.
7. Newell, J.W., Seo, N.S., Enns, G.M., McCraken, M., Mantovani, J.F. and Freeze, H.H. 2003. Congenital disorder of glycosylation 1C in patients of Indian origin. Mol. Genet. Metab. 79: 221-228.
8. Sun, L., Eklund, E.A., Van Hove, J.L., Freeze, H.H. and Thomas, J.A. 2005. Clinical and molecular characterization of the first adult congenital disorder of glycosylation (CDG) type 1C patient. Am. J. Med. Genet. A 137: 22-26.
9. Uchimura, S., Sugiyama, M. and Nikawa, J. 2005. Effects of N-glycosylation and inositol on the ER stress response in yeast *Saccharomyces cerevisiae*. Biosci. Biotechnol. Biochem. 69: 1274-1280.

CHROMOSOMAL LOCATION

Genetic locus: ALG6 (human) mapping to 1p31.3; Alg6 (mouse) mapping to 4 C6.

SOURCE

ALG6 (Z164) is a mouse monoclonal antibody raised against recombinant ALG6 of human origin.

PRODUCT

Each vial contains 100 µg IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

ALG6 (Z164) is recommended for detection of ALG6 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)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for ALG6 siRNA (h): sc-88385, ALG6 siRNA (m): sc-141016, ALG6 shRNA Plasmid (h): sc-88385-SH, ALG6 shRNA Plasmid (m): sc-141016-SH, ALG6 shRNA (h) Lentiviral Particles: sc-88385-V and ALG6 shRNA (m) Lentiviral Particles: sc-141016-V.

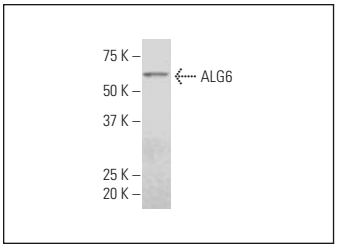
Molecular Weight of ALG6: 58 kDa.

Positive Controls: HeLa nuclear extract: sc-2120.

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).

DATA



ALG6 (Z164): sc-100506. Western blot analysis of ALG6 expression in HeLa nuclear extract.

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.