SANTA CRUZ BIOTECHNOLOGY, INC.

AGL (N-16): sc-161316

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

AGL (amylo-1,6-glucosidase, 4-α-glucotransferase), also known as GDE (glycogen debranching enzyme), is a 1,532 amino acid protein that exists as 3 alternatively spliced isoforms which are expressed in kidney, liver, heart and muscle in an isoform-specific manner. Exhibiting multifunctional enzyme capabilities, AGL contains two catalytic active sites, one of which acts as an 4-α-glucotransferase and the other of which acts as an amylo-1,6-glucosidase during glycogen degradation. Defects in the gene encoding AGL are the cause of glycogen storage disease type 3 (GSD3), also known as Forbes disease. GSD3 is a metabolic disorder that is characterized by the presence of abnormal glycogen due to a lack of AGL activity. Symptoms of GSD3 generally include hypoglycemia, variable myopathy, hepatomegaly and short stature.

REFERENCES

1. Ding, J.H., de Barsy, T., Brown, B.I., Coleman, R.A. and Chen, Y.T. 1990. Immunoblot analyses of glycogen debranching enzyme in different subtypes of glycogen storage disease type III. J. Pediatr. 116: 95-100.
2. Yang, B.Z., Ding, J.H., Enghild, J.J., Bao, Y. and Chen, Y.T. 1992. Molecular cloning and nucleotide sequence of cDNA encoding human muscle glycogen debranching enzyme. J. Biol. Chem. 267: 9294-9299.
3. Shen, J., Bao, Y., Liu, H.M., Lee, P., Leonard, J.V. and Chen, Y.T. 1996. Mutations in exon 3 of the glycogen debranching enzyme gene are associated with glycogen storage disease type III that is differentially expressed in liver and muscle. J. Clin. Invest. 98: 352-357.
4. Orho, M., Bosshard, N.U., Buist, N.R., Gitzelmann, R., Aynsley-Green, A., Blümel, P., Gannon, M.C., Nuttall, F.Q. and Groop, L.C. 1998. Mutations in the liver glycogen synthase gene in children with hypoglycemia due to glycogen storage disease type 0. J. Clin. Invest. 102: 507-515.
5. Horinishi, A., Okubo, M., Tang, N.L., Hui, J., To, K.F., Mabuchi, T., Okada, T., Mabuchi, H. and Murase, T. 2002. Mutational and haplotype analysis of AGL in patients with glycogen storage disease type III. J. Hum. Genet. 47: 55-59.
6. Sakoda, H., Fujishiro, M., Fujio, J., Shojima, N., Ogihara, T., Kushiyama, A., Fukushima, Y., Anai, M., Ono, H., Kikuchi, M., Horike, N., Viana, A.Y., Uchijima, Y., Kurihara, H. and Asano, T. 2005. Glycogen debranching enzyme association with β-subunit regulates AMP-activated protein kinase activity. Am. J. Physiol. Endocrinol. Metab. 289: E474-E481.
7. Endo, Y., Horinishi, A., Vorgerd, M., Aoyama, Y., Ebara, T., Murase, T., Odawara, M., Podskarbi, T., Shin, Y.S. and Okubo, M. 2006. Molecular analysis of the AGL gene: heterogeneity of mutations in patients with glycogen storage disease type III from Germany, Canada, Afghanistan, Iran, and Turkey. J. Hum. Genet. 51: 958-963.
8. Lucchiari, S., Santoro, D., Pagliarani, S. and Comi, G.P. 2007. Clinical, biochemical and genetic features of glycogen debranching enzyme deficiency. Acta Myol. 26: 72-74.
9. Cheng, A., Zhang, M., Gentry, M.S., Worby, C.A., Dixon, J.E. and Saltiel, A.R. 2007. A role for AGL ubiquitination in the glycogen storage disorders of Lafora and Cori's disease. Genes Dev. 21: 2399-2409.

CHROMOSOMAL LOCATION

Genetic locus: AGL (human) mapping to 1p21.2; Agl (mouse) mapping to 3 G1.

SOURCE

AGL (N-16) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of AGL 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-161316 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

AGL (N-16) is recommended for detection of AGL 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).

AGL (N-16) is also recommended for detection of AGL in additional species, including equine, canine and bovine.

Suitable for use as control antibody for AGL siRNA (h): sc-88368, AGL siRNA (m): sc-140904, AGL shRNA Plasmid (h): sc-88368-SH, AGL shRNA Plasmid (m): sc-140904-SH, AGL shRNA (h) Lentiviral Particles: sc-88368-V and AGL shRNA (m) Lentiviral Particles: sc-140904-V.

Molecular Weight of AGL: 160 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.

RESEARCH USE

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

PROTOCOLS

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