SANTA CRUZ BIOTECHNOLOGY, INC.

LYAG (Q-17): sc-49441

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Lysosomal α-glucosidase (LYAG), also designated acid α-glucosidase or acid maltase, is essential for the degradation of glycogen to glucose in lysosomes. LYAG is a protein belonging to the glycosyl hydrolase 31 family and resides soley in the lysosome. After translation, LYAG undergoes proteolytic processing to form two lengths of lysosomal a-glucosidase, and both N-terminal and C-terminal processing occur. Conduritol B epoxide (CBE) is a competitive inhibitor of LYAG. Defects in GAA, the gene encoding for LYAG, may cause Pompe disease, an autosomal recessive disorder characterized by cardiorespiratory insufficiency and glycogen accumulation in muscle tissues, causing muscular weakness. Mutations on the LYAG gene also cause glycogen storage disease II (GSD-II).

REFERENCES

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2. Winkel, L.P., et al. 2005. The natural course of non-classic Pompe's disease; a review of 225 published cases. J. Neurol. 252: 875-884.
3. Umapathysivam, K., et al. 2005. Correlation of acid α-glucosidase and glycogen content in skin fibroblasts with age of onset in Pompe disease. Clin. Chim. Acta 361: 191-198.
4. Sharma, M.C., et al. 2005. Delayed or late-onset type II glycogenosis with globular inclusions. Acta Neuropathol. 110: 151-157.
5. Mah, C., et al. 2005. Sustained correction of glycogen storage disease type II using adeno-associated virus serotype 1 vectors. Gene Ther. 12: 1405-1409.
6. Cresawn, K.O., et al. 2005. Impact of humoral immune response on distribution and efficacy of recombinant adeno-associated virus-derived acid α-glucosidase in a model of glycogen storage disease type II. Hum. Gene Ther. 16: 68-80.
7. Klinge, L., et al. 2005. Safety and efficacy of recombinant acid α-glucosidase (rhGAA) in patients with classical infantile Pompe disease: results of a phase II clinical trial. Neuromuscul. Disord. 15: 24-31.

CHROMOSOMAL LOCATION

Genetic locus: GAA (human) mapping to 17q25.3; Gaa (mouse) mapping to 11 E2.

SOURCE

LYAG (Q-17) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of LYAG 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-49441 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

LYAG (Q-17) is recommended for detection of precursor and mature lysosomal α-glucosidase (LYAG) 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).

LYAG (Q-17) is also recommended for detection of precursor and mature lysosomal α-glucosidase (LYAG) in additional species, including equine and bovine.

Suitable for use as control antibody for LYAG siRNA (h): sc-60974, LYAG siRNA (m): sc-60975, LYAG shRNA Plasmid (h): sc-60974-SH, LYAG shRNA Plasmid (m): sc-60975-SH, LYAG shRNA (h) Lentiviral Particles: sc-60974-V and LYAG shRNA (m) Lentiviral Particles: sc-60975-V.

Molecular Weight of LYAG cleavage fragments: 70/76 kDa.

Molecular Weight of LYAG: 110 kDa.

Positive Controls: COLO 320DM cell lysate: sc-2226.

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.