SANTA CRUZ BIOTECHNOLOGY, INC.

LYAG (M-63): sc-67358

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 α-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

1. Sohar, N., et al. 2005. Lysosomal enzyme activities: new potential markers for Sjogren's syndrome Clin. Biochem. 38:1120-1126.
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.

CHROMOSOMAL LOCATION

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

SOURCE

LYAG (M-63) is a rabbit polyclonal antibody raised against amino acids 129-191 mapping near the N-terminus of LYAG of mouse origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

LYAG (M-63) is recommended for detection of precursor and mature lysosomal α-glucosidase (LYAG) of mouse, rat and, to a lesser extent, 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)], 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).

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.

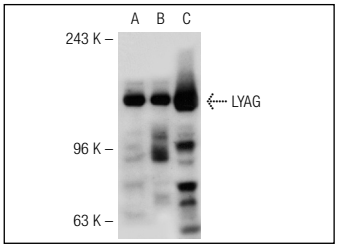
Molecular Weight of LYAG: 110 kDa.

Positive Controls: LYAG (m): 293T Lysate: sc-125567.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



LYAG (M-63): sc-67358. Western blot analysis of LYAG expression in non-transfected 293T: sc-117752 (**A**), mouse LYAG transfected 293T: sc-125567 (**B**) and COLO 320DM (**C**) whole cell lysates.

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.