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Produktinformation



Forschungsprodukte & Biochemikalien



Zellkultur & Verbrauchsmaterial



Diagnostik & molekulare Diagnostik



Laborgeräte & Service

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Lieferung & Zahlungsart

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Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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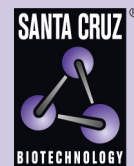
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LYAG (h): 293T Lysate: sc-115825

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 solely 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. Conduiritol 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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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.
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8. 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.

PRODUCT

LYAG (h): 293T Lysate represents a lysate of human LYAG transfected 293T cells and is provided as 100 μ g protein in 200 μ l SDS-PAGE buffer.

APPLICATIONS

LYAG (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive LYAG antibodies. Recommended use: 10-20 μ l per lane.

Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. 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 for detailed protocols and support products.