SANTA CRUZ BIOTECHNOLOGY, INC.

LYAG siRNA (m): sc-60975

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 Sjögren's syndrome. Clin. Biochem. 38: 1120-1126.
2. Winkel, L.P., et al. 2005. The natural course of non-classic Pompe 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. Anneser, J.M., et al. 2005. Mutations in the acid α-glucosidase gene *(M. Pompe)* in a patient with an unusual phenotype. Neurology 64: 368-370.

CHROMOSOMAL LOCATION

Genetic locus: Gaa (mouse) mapping to 11 E2.

PRODUCT

LYAG siRNA (m) is a pool of 3 target-specific 19-25 nt siRNAs designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10 µM solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see LYAG shRNA Plasmid (m): sc-60975-SH and LYAG shRNA (m) Lentiviral Particles: sc-60975-V as alternate gene silencing products.

For independent verification of LYAG (m) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-60975A, sc-60975B and sc-60975C.

RESEARCH USE

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

STORAGE AND RESUSPENSION

Store lyophilized siRNA duplex at -20° C with desiccant. Stable for at least one year from the date of shipment. Once resuspended, store at -20° C, avoid contact with RNAses and repeated freeze thaw cycles.

Resuspend lyophilized siRNA duplex in 330 µl of the RNAse-free water provided. Resuspension of the siRNA duplex in 330 µl of RNAse-free water makes a 10 µM solution in a 10 µM Tris-HCL, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

LYAG siRNA (m) is recommended for the inhibition of LYAG expression in mouse cells.

SUPPORT REAGENTS

For optimal siRNA transfection efficiency, Santa Cruz Biotechnology's siRNA Transfection Reagent: sc-29528 (0.3 ml), siRNA Transfection Medium: sc-36868 (20 ml) and siRNA Dilution Buffer: sc-29527 (1.5 ml) are recommended. Control siRNAs or Fluorescein Conjugated Control siRNAs are available as 10 µM in 66 µl. Each contain a scrambled sequence that will not lead to the specific degradation of any known cellular mRNA. Fluorescein Conjugated Control siRNAs include: sc-36869, sc-44239, sc-44240 and sc-44241. Control siRNAs include: sc-37007, sc-44230, sc-44231, sc-44232, sc-44233, sc-44234, sc-44235, sc-44236, sc-44237 and sc-44238.

GENE EXPRESSION MONITORING

LYAG (G-7): sc-373745 is recommended as a control antibody for monitoring of LYAG gene expression knockdown by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) or immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

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) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor LYAG gene expression knockdown using RT-PCR Primer: LYAG (m)-PR: sc-60975-PR (20 µl, 580 bp). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

PROTOCOLS

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