SANTA CRUZ BIOTECHNOLOGY, INC.

GBE1 siRNA (h): sc-78413

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

GBE1 (glucan (1,4-α-), branching enzyme 1) is a 702 amino acid protein that is expressed at high levels in muscle and liver and is involved in glycogen biosynthesis. Existing as a monomer, GBE1 catalyzes the transfer of α-1,4-linked glucosyl units from the outer end of a glycogen chain to an α-1,6 position on a neighboring glycogen chain and, via this catalytic activity, plays an essential role in glycogen accumulation. Defects in the gene encoding GBE1 are the cause of glycogen storage disease type 4 (GSD4) and adult polyglucosan body disease (APBD), the first of which is a metabolic disorder that is associated with the accumulation of polysaccharides and is characterized by liver disease during childhood. Unlike GSD4, APBD is a late-onset disorder that affects the central and peripheral nervous systems and is characterized by cognitive impairment, pyramidal tetraparesis and peripheral neuropathy.

REFERENCES

1. Brown, D.H., et al. 1983. Studies of the residual glycogen branching enzyme activity present in human skin fibroblasts from patients with type IV glycogen storage disease. Biochem. Biophys. Res. Commun. 111: 636-643.
2. Bao, Y., et al. 1996. Hepatic and neuromuscular forms of glycogen storage disease type IV caused by mutations in the same glycogen-branching enzyme gene. J. Clin. Invest. 97: 941-948.
3. Alegria, A., et al. 1999. Glycogen storage disease type IV presenting as hydrops fetalis. J. Inherit. Metab. Dis. 22: 330-332.
4. Chan, Y.J., et al. 1999. Glycogen storage disease type IV: a case report. Zhonghua Yi Xue Za Zhi 62: 743-747.
5. Ziemssen, F., et al. 2000. Novel missense mutations in the glycogen-branching enzyme gene in adult polyglucosan body disease. Ann. Neurol. 47: 536-540.
6. Bruno, C., et al. 2004. Clinical and genetic heterogeneity of branching enzyme deficiency (glycogenosis type IV). Neurology 63: 1053-1058.

CHROMOSOMAL LOCATION

Genetic locus: GBE1 (human) mapping to 3p12.2.

PRODUCT

GBE1 siRNA (h) 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 GBE1 shRNA Plasmid (h): sc-78413-SH and GBE1 shRNA (h) Lentiviral Particles: sc-78413-V as alternate gene silencing products.

For independent verification of GBE1 (h) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-78413A, sc-78413B and sc-78413C.

PROTOCOLS

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

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

GBE1 siRNA (h) is recommended for the inhibition of GBE1 expression in human 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.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor GBE1 gene expression knockdown using RT-PCR Primer: GBE1 (h)-PR: sc-78413-PR (20 µl). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

RESEARCH USE

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