

GLUD1 (h2): 293T Lysate: sc-115824

BACKGROUND

GLUD1 (glutamate dehydrogenase 1), also known as GDH, GDH1 or GLUD, is a 558 amino acid mitochondrial matrix enzyme belonging to the Glu/Leu/Phe/Val dehydrogenases family. Existing as a homohexamer, GLUD1 catalyzes the oxidative deamination of glutamate to α -ketoglutarate and ammonia. GLUD1 is critical for regulating amino acid-induced Insulin secretion and is allosterically activated by ADP and inhibited by GTP and ATP. Mutations in the gene encoding GLUD1 causes hyperInsulinism-hyperammonemia syndrome (HHS), which is an inherited condition characterized by high Insulin and ammonia levels in the blood. GLUD1 may also be involved in learning and memory reactions by increasing the turnover of the excitatory neurotransmitter glutamate.

REFERENCES

1. Stanley, C.A., et al. 2000. Molecular basis and characterization of the hyperInsulinism/hyperammonemia syndrome: predominance of mutations in exons 11 and 12 of the glutamate dehydrogenase gene. *HI/HA Contributing Investigators. Diabetes* 49: 667-673.
2. Tanizawa, Y., et al. 2002. Unregulated elevation of glutamate dehydrogenase activity induces glutamine-stimulated Insulin secretion: identification and characterization of a GLUD1 gene mutation and Insulin secretion studies with MIN6 cells overexpressing the mutant glutamate dehydrogenase. *Diabetes* 51: 712-717.
3. de Lonlay, P., et al. 2005. Dominantly inherited hyperInsulinaemic hypoglycaemia. *J. Inherit. Metab. Dis.* 28: 267-276.
4. Mastorodemos, V., et al. 2009. Human GLUD1 and GLUD2 glutamate dehydrogenase localize to mitochondria and endoplasmic reticulum. *Biochem. Cell Biol.* 87: 505-516.
5. Kanavouras, K., et al. 2009. Mutations in human GLUD2 glutamate dehydrogenase affecting basal activity and regulation. *J. Neurochem.* 109: 167-173.
6. Kapoor, R.R., et al. 2009. Advances in the diagnosis and management of hyperInsulinemic hypoglycemia. *Nat. Clin. Pract. Endocrinol. Metab.* 5: 101-112.
7. Zaganas, I., et al. 2009. The human GLUD2 glutamate dehydrogenase: localization and functional aspects. *Neurochem. Int.* 55: 52-63.
8. Paji, T., et al. 2009. Glutamate dehydrogenase activity in lymphocytes of B-cell chronic lymphocytic leukaemia patients. *Clin. Biochem.* 42: 1677-1684.
9. Kapoor, R., et al. 2009. HyperInsulinism-hyperammonemia (HI/HA) syndrome: novel mutations in the GLUD1 gene and genotype-phenotype correlations. *Eur. J. Endocrinol.* 161: 731-735.

CHROMOSOMAL LOCATION

Genetic locus: GLUD1 (human) mapping to 10q23.2.

PRODUCT

GLUD1 (h2): 293T Lysate represents a lysate of human GLUD1 transfected 293T cells and is provided as 100 μ g protein in 200 μ l SDS-PAGE buffer.

APPLICATIONS

GLUD1 (h2): 293T Lysate is suitable as a Western Blotting positive control for human reactive GLUD1 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.