

GSS (h2): 293T Lysate: sc-170509

BACKGROUND

GSS (Glutathione synthetase) is a 474 amino acid protein encoded by the gene located at chromosome 20q11.22. GSS consists of three loops projecting from an antiparallel β -sheet, a parallel β -sheet and a lid of anti-parallel sheets, which provide access to the ATP-binding site. Although Southern blot and gene analysis suggest that GSS may be the only member of a unique family, the crystal structure indicates that GSS belongs to the ATP-GRASP superfamily. GSS is expressed in hemocytes and nucleated cells including the brain. GSS occurs as a homodimer. There are two steps in the production of Glutathione, beginning with γ -GCS and ending with GSS. In an ATP-dependent reaction, GSS produces Glutathione from γ -glutamylcysteine and glycine precursors. Partial hepatectomy, diethyl maleate, buthionine sulfoximine, tert-butylhydroquinone and thioacetamide increase the expression of GSS, which causes an increase in Glutathione levels. 5-oxoprolinuria (pyroglutamic aciduria), an inherited autosomal recessive disorder, is caused by GSS deficiencies, which leads to central nervous system damage, hemolytic anemia, metabolic acidosis and urinary excretion of 5-oxoproline. A missense mutation in the gene encoding GSS leads to a GSS deficiency restricted to erythrocytes, which causes only hemolytic anemia.

REFERENCES

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2. Gali, R.R. and Board, P.G. 1995. Sequencing and expression of a cDNA for human Glutathione synthetase. *Biochem. J.* 310: 353-358
3. Shi, Z.Z., Habib, G.M., Rhead, W.J., Gahl, W.A., He, X., Sazer, S. and Lieberman, M.W. 1996. Mutations in the Glutathione synthetase gene cause 5-oxoprolinuria. *Nat. Genet.* 14: 361-365.
4. Polekhina, G., Board, P.G., Gali, R.R., Rossjohn, J. and Parker, M.W. 1999. Molecular basis of Glutathione synthetase deficiency and a rare gene permutation event. *EMBO J.* 18: 3204-3213.
5. Huang, Z.A., Yang, H., Chen, C., Zeng, Z. and Lu, S.C. 2000. Inducers of γ -glutamylcysteine synthetase and their effects on Glutathione synthetase expression. *Biochim. Biophys. Acta* 1493: 48-55.

CHROMOSOMAL LOCATION

Genetic locus: GSS (human) mapping to 20q11.22.

PRODUCT

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

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.

PROTOCOLS

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

APPLICATIONS

GSS (h2): 293T Lysate is suitable as a Western Blotting positive control for human reactive GSS antibodies. Recommended use: 10-20 μ l per lane.

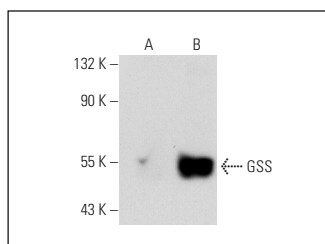
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

GSS (H-7): sc-166882 is recommended as a positive control antibody for Western Blot analysis of enhanced human GSS expression in GSS transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended:
1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ 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.

DATA



GSS (H-7): sc-166882. Western blot analysis of GSS expression in non-transfected: sc-117752 (A) and human GSS transfected: sc-170509 (B) 293T whole cell lysates.

RESEARCH USE

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