



# SZABO SCANDIC

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- Gefahrgutzuschlag
- Expressversand

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# 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  $\beta$ -sheet, a parallel  $\beta$ -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  $\gamma$ -GCS and ending with GSS. In an ATP-dependent reaction, GSS produces Glutathione from  $\gamma$ -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

1. Webb, G.C., Vaska, V.L., Gali, R.R., Ford, J.H. and Board, P.G. 1995. The gene encoding human Glutathione synthetase (GSS) maps to the long arm of chromosome 20 at band 11.2. *Genomics* 30: 617-619.
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  $\gamma$ -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  $\mu$ g protein in 200  $\mu$ 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](http://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  $\mu$ l per lane.

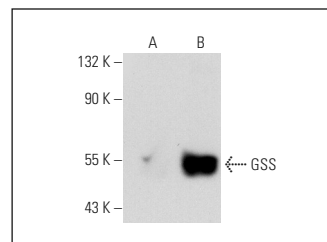
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

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 $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  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.