

GlyRS (m): 293T Lysate: sc-120534

BACKGROUND

The fidelity of protein synthesis requires efficient discrimination of amino acid substrates by aminoacyl-tRNA synthetases. Proteins belonging to this family function to catalyze the aminoacylation of tRNAs by their corresponding amino acids, thus linking amino acids with tRNA-contained nucleotide triplets. GlyRS (Glycyl-tRNA synthetase), also known as Glycine-tRNA ligase, is a 739 amino acid class II synthetase that is widely expressed, including in the brain and spinal cord. Defects in the gene encoding GlyRS is the cause of Charcot-Marie-Tooth disease type 2D (CMT2D), which is an autosomal dominant inherited disease characterized by severe weakness, atrophy and absence of deep tendon reflexes in the upper extremities. Defects in the GlyRS gene is also the cause of distal hereditary muscular neuropathy type V (HMN5), a disease similar to CMT2D, though the distal sensory involvement is less severe in HMN5 patients.

REFERENCES

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2. Williams, J., et al. 1995. Cloning, sequencing and bacterial expression of human glycine tRNA synthetase. *Nucleic Acids Res.* 23: 1307-1310.
3. Antonellis, A., et al. 2003. Glycyl tRNA synthetase mutations in Charcot-Marie-Tooth disease type 2D and distal spinal muscular atrophy type V. *Am. J. Hum. Genet.* 72: 1293-1299.
4. Antonellis, A., et al. 2006. Functional analyses of glycyl-tRNA synthetase mutations suggest a key role for tRNA-charging enzymes in peripheral axons. *J. Neurosci.* 26: 10397-10406.
5. James, P.A., et al. 2006. Severe childhood SMA and axonal CMT due to anticodon binding domain mutations in the GARS gene. *Neurology* 67: 1710-1712.
6. Seburn, K.L., et al. 2006. An active dominant mutation of glycyl-tRNA synthetase causes neuropathy in a Charcot-Marie-Tooth 2D mouse model. *Neuron* 51: 715-726.

CHROMOSOMAL LOCATION

Genetic locus: Gars (mouse) mapping to 6 B3.

PRODUCT

GlyRS (m): 293T Lysate represents a lysate of mouse GlyRS transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

GlyRS (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive GlyRS antibodies. Recommended use: 10-20 µl per lane.

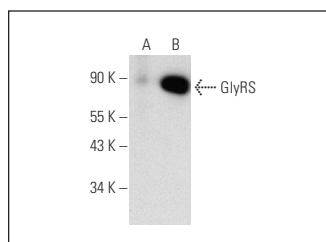
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

GlyRS (H-1): sc-271778 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse GlyRS expression in GlyRS 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-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.

DATA



GlyRS (H-1): sc-271778. Western blot analysis of GlyRS expression in non-transfected: sc-117752 (A) and mouse GlyRS transfected: sc-120534 (B) 293T whole cell lysates.

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.