



# SZABO SCANDIC

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## Produktinformation



Forschungsprodukte & Biochemikalien



Zellkultur & Verbrauchsmaterial



Diagnostik & molekulare Diagnostik



Laborgeräte & Service

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### Lieferung & Zahlungsart

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### Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

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# SPR (m): 293T Lysate: sc-123752

## BACKGROUND

SPR, also known as sepiapterin reductase, is a homodimeric cytoplasmic protein that belongs to the sepiapterin reductase family. SPR functions as an NADH-dependent aldo-keto reductase and specifically catalyzes the reduction of pteridine derivatives. In addition, SPR plays an important role in tetrahydrobiopterin (BH4) biosynthesis, catalyzing the final reduction step of the synthesis pathway. BH4 is an essential cofactor for the hydroxylation of the aromatic amino acids (tryptophan, tyrosine and phenylalanine) and is required for proper dopamine synthesis. Mutations in the gene encoding SPR can cause sepiapterin reductase deficiency, a monoamine neurotransmitter deficiency without hyperphenylalaninemia. Sepiapterin reductase deficiency interferes with BH4 synthesis, resulting in DOPA-responsive dystonia and a variety of other human diseases. In addition, SPR mRNA expression is increased in the brain of Parkinson's Disease (PD) patients, suggesting that SPR may play a role in PD.

## REFERENCES

- Auerbach, G., et al. 1997. The 1.25 Å crystal structure of sepiapterin reductase reveals its binding mode to pterins and brain neurotransmitters. *EMBO J.* 16: 7219-7230.
- Blau, N., et al. 2001. Tetrahydrobiopterin deficiencies without hyperphenylalaninemia: diagnosis and genetics of dopa-responsive dystonia and sepiapterin reductase deficiency. *Mol. Genet. Metab.* 74: 172-185.
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- Friedman, J., et al. 2006. Dopa-responsive hypersomnia and mixed movement disorder due to sepiapterin reductase deficiency. *Neurology* 67: 2032-2035.
- Echenne, B., et al. 2006. Sepiapterin reductase deficiency: clinical presentation and evaluation of long-term therapy. *Pediatr. Neurol.* 35: 308-313.
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- Kim, H.L., et al. 2007. Tetrahydropteridine deficiency impairs mitochondrial function in Dictyostelium discoideum Ax2. *FEBS Lett.* 581: 5430-5434.
- Takazawa, C., et al. 2008. A brain-specific decrease of the tyrosine hydroxylase protein in sepiapterin reductase-null mice—as a mouse model for Parkinson's disease. *Biochem. Biophys. Res. Commun.* 367: 787-792.

## CHROMOSOMAL LOCATION

Genetic locus: Spr (mouse) mapping to 6 C3.

## PRODUCT

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

## APPLICATIONS

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

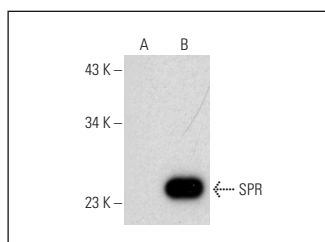
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

SPR (H-12): sc-398126 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse SPR expression in SPR transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

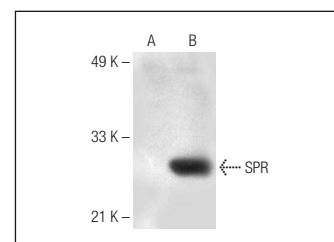
## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-mouse IgG-HRP: sc-2005 (dilution range: 1:2000-1:32,000) or Cruz Marker™ compatible goat anti-mouse IgG-HRP: sc-2031 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048.

## DATA



SPR (H-12): sc-398126. Western blot analysis of SPR expression in non-transfected: sc-117752 (A) and mouse SPR transfected: sc-123752 (B) 293T whole cell lysates.



SPR (G-14): sc-169414. Western blot analysis of SPR expression in non-transfected: sc-117752 (A) and mouse SPR transfected: sc-123752 (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](http://www.scbt.com) or our catalog for detailed protocols and support products.