

SPR (H-12): sc-398126

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

1. 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.
2. 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.
3. Ikemoto, K., et al. 2002. Localization of sepiapterin reductase in the human brain. *Brain Res.* 954: 237-246.
4. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 182125. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
5. Friedman, J., et al. 2006. Dopa-responsive hypersomnia and mixed movement disorder due to sepiapterin reductase deficiency. *Neurology* 67: 2032-2035.

CHROMOSOMAL LOCATION

Genetic locus: SPR (human) mapping to 2p13.2; Spr (mouse) mapping to 6 C3.

SOURCE

SPR (H-12) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 191-214 near the C-terminus of SPR of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2b} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-398126 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

APPLICATIONS

SPR (H-12) is recommended for detection of SPR of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

SPR (H-12) is also recommended for detection of SPR in additional species, including equine and canine.

Suitable for use as control antibody for SPR siRNA (h): sc-94595, SPR siRNA (m): sc-153782, SPR shRNA Plasmid (h): sc-94595-SH, SPR shRNA Plasmid (m): sc-153782-SH, SPR shRNA (h) Lentiviral Particles: sc-94595-V and SPR shRNA (m) Lentiviral Particles: sc-153782-V.

Molecular Weight of SPR monomer: 30 kDa.

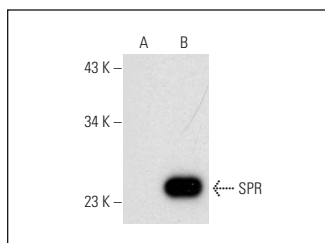
Molecular Weight of SPR dimer: 56 kDa.

Positive Controls: SPR (m): 293T Lysate: sc-123752.

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. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

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.

SELECT PRODUCT CITATIONS

1. Shin, N., et al. 2019. Uncoupled endothelial nitric oxide synthase enhances p-Tau in chronic traumatic encephalopathy mouse model. *Antioxid. Redox Signal.* 30: 1601-1620.

RESEARCH USE

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