

SPR (A-11): sc-514777

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/>

CHROMOSOMAL LOCATION

Genetic locus: SPR (human) mapping to 2p13.2.

SOURCE

SPR (A-11) is a mouse monoclonal antibody raised against amino acids 1-261 representing full length SPR of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

SPR (A-11) is available conjugated to agarose (sc-514777 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-514777 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-514777 PE), fluorescein (sc-514777 FITC), Alexa Fluor® 488 (sc-514777 AF488), Alexa Fluor® 546 (sc-514777 AF546), Alexa Fluor® 594 (sc-514777 AF594) or Alexa Fluor® 647 (sc-514777 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-514777 AF680) or Alexa Fluor® 790 (sc-514777 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

SPR (A-11) is recommended for detection of SPR of 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).

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

Molecular Weight of SPR monomer: 30 kDa.

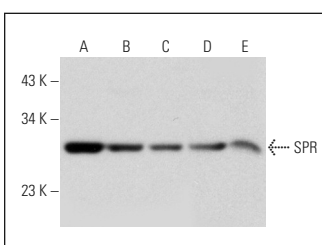
Molecular Weight of SPR dimer: 56 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, Caki-1 cell lysate: sc-2224 or K-562 whole cell lysate: sc-2203.

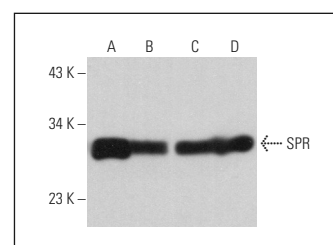
RECOMMENDED SUPPORT REAGENTS

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 (A-11): sc-514777. Western blot analysis of SPR expression in Hep G2 (A), Caki-1 (B), K-562 (C) and THP-1 (D) whole cell lysates and human skeletal muscle tissue extract (E).



SPR (A-11): sc-514777. Western blot analysis of SPR expression in A549 (A), MCF7 (B), Hep G2 (C) and Caki-1 (D) whole cell lysates.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. 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.