10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

SPR Rabbit pAb

Catalog # AP97542

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P35270
Other Accession	P35270
Reactivity	Rat, Human, Mouse
Predicted	Pig, Rabbit, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	28048
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Sepiapterin reductase
Epitope Specificity	101-200/261
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the sepiapterin reductase family.
SUBUNIT	Homodimer.
Post-translational modifications	In vitro phosphorylation of Ser-213 by CaMK2 does not change kinetic parameters.
DISEASE	Defects in SPR are the cause of dystonia DOPA-responsive due to sepiapterin reductase deficiency (DRDSPRD) [MIM:612716]. In the majority of cases, patients manifest progressive psychomotor retardation, dystonia and spasticity. Cognitive anomalies are also often present. The disease is due to severe dopamine and serotonin deficiencies in the central nervous system caused by a defect in BH4 synthesis. Dystonia is defined by the presence of sustained involuntary muscle contractions, often leading to abnormal postures.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes an aldo-keto reductase that catalyzes the NADPH-dependent reduction of pteridine derivatives and is important in the biosynthesis of tetrahydrobiopterin (BH4). Mutations in this gene result in DOPA-responsive dystonia due to sepiaterin reductase deficiency. A pseudogene has been identified on chromosome 1. [provided by RefSeq, Jul 2008]

Additional Information

Gene ID	6697
Other Names	SPRE_HUMAN; SPR; 1.1.1.153; sepiapterin reductase; sepiapterin reductase (7, 8-dihydrobiopterin:NADP+ oxidoreductase); short chain dehydrogenase/reductase family 38C, member 1; Sepiapterin reductase

	(L-erythro-7, 8-dihydrobiopterin forming)
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	SPR
Function	Catalyzes the final one or two reductions in tetra- hydrobiopterin biosynthesis to form 5,6,7,8-tetrahydrobiopterin.
Cellular Location	Cytoplasm.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.