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Rabbit Anti-SPR antibody

SL11784R

Product Name:	SPR
Chinese Name:	墨蝶蛉还原酶抗体
Alias:	SDR38C1; Sepiapterin reductase (7,8 dihydrobiopterin:NADP+ oxidoreductase); Sepiapterin reductase; Short chain dehydrogenase/reductase family 38C, member 1; SPR; SPRE_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Pig,Horse,Rabbit,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	28kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Sepiapterin reductase:101-200/261
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	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]

Function:
Catalyzes the final one or two reductions in tetra-hydrobiopterin biosynthesis to form 5,6,7,8-tetrahydrobiopterin.

Subunit:
Homodimer.

Subcellular Location:
Cytoplasm.

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.

Similarity:
Belongs to the sepiapterin reductase family.

SWISS:
P35270

Gene ID:
6697

Database links:

Entrez Gene: 6697 Human

Omim: 182125 Human

SwissProt: P35270 Human

Unigene: 301540 Human

Important Note:
This product as supplied is intended for research use only, not for use in human,

	therapeutic or diagnostic applications.