abcam

Product datasheet

Recombinant Human SPR protein ab86445

1 Image

Description

Product name Recombinant Human SPR protein

Purity > 95 % SDS-PAGE.

ab86445 is purified using conventional chromatography techniques.

Expression system Escherichia coli

Protein length Full length protein

Animal free No

Nature Recombinant

Species Human

Sequence MGSSHHHHHH SSGLVPRGSH MEGGLGRAVC

LLTGASRGFG RTLAPLLASL LSPGSVLVLS
ARNDEALRQL EAELGAERSG LRVVRVPADL
GAEAGLQQLL GALRELPRPK GLQRLLLINN
AGSLGDVSKG FVDLSDSTQV NNYWALNLTS
MLCLTSSVLK AFPDSPGLNR TVVNISSLCA
LQPFKGWALY CAGKAARDML FQVLALEEPN
VRVLNYAPGP LDTDMQQLAR ETSVDPDMRK
GLQELKAKGK LVDCKVSAQK LLSLLEKDEF

KSGAHVDFYD K

Specifications

Our **Abpromise guarantee** covers the use of **ab86445** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications SDS-PAGE

Form Liquid

Additional notes Endotoxin Level: < 1.0 EU per 1 µg of protein (determined by LAL method).

Preparation and Storage

Stability and Storage Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C or -

80°C. Avoid freeze / thaw cycle.

pH: 8.00

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General Info

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

tetrahydrobiopterin.

Involvement in 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.

Sequence similarities Belongs to the sepiapterin reductase family.

Post-translational modifications

In vitro phosphorylation of Ser-213 by CaMK2 does not change kinetic parameters.

Cellular localization Cytoplasm.

Images



15% SDS-PAGE showing ab86445 at approximately 30.2kDa ($3\mu g$).

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