PDSS1 (C-8): sc-518218



The Power to Question

BACKGROUND

PDSS1, decaprenyl-diphosphate synthase subunit 1, is a magnesium binding peptide that belongs to the FPP/GGPP synthetase family. Forming a heterotetramer that consists of 2 DPS1/TPRT and 2 DLP1 subunits, Decaprenyl-diphosphate synthase functions to supply decaprenyl diphosphate, which is the precursor for the side chains of the isoprenoid quinones ubiquinone-10. Limited expression or defects of PDSS1 can lead to a coenzyme Ω_{10} deficiency which can be manifested by several phenotypes. Coenzyme Ω_{10} (Co Ω_{10}) deficiencies can lead to reduced ATP synthesis and result in marked cerebellar atrophy and pure myopathy. $\text{Co}\Omega_{10}$ deficiencies has also been associated with reversible renal diseases and infantile multisystemic and cerebellar ataxia.

REFERENCES

- 1. Park, Y.C., et al. 2005. Batch and fed-batch production of coenzyme Ω_{10} in recombinant *Escherichia coli* containing the decaprenyl diphosphate synthase gene from *Gluconobacter suboxydans*. Appl. Microbiol. Biotechnol. 67: 192-196.
- Saiki, R., et al. 2005. Characterization of solanesyl and decaprenyl diphosphate synthases in mice and humans. FEBS J. 272: 5606-5622.
- 3. Takahashi, S., et al. 2006. Metabolic engineering of coenzyme Q by modification of isoprenoid side chain in plant. FEBS Lett. 580: 955-959.
- 4. Zahiri, H.S., et al. 2006. Coenzyme Q₁₀ production in recombinant Escherichia coli strains engineered with a heterologous decaprenyl diphosphate synthase gene and foreign mevalonate pathway. Metab. Eng. 8: 406-416.
- Mollet, J., et al. 2007. Prenyldiphosphate synthase, subunit 1 (PDSS1) and OH-benzoate polyprenyltransferase (COQ2) mutations in ubiquinone deficiency and oxidative phosphorylation disorders. J. Clin. Invest. 117: 765-772.

CHROMOSOMAL LOCATION

Genetic locus: PDSS1 (human) mapping to 10p12.1.

SOURCE

PDSS1 (C-8) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 174-196 of PDSS1 of human origin.

PRODUCT

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

PDSS1 (C-8) is available conjugated to agarose (sc-518218 AC), 500 μ g/ 0.25 ml agarose in 1 ml, for IP; to HRP (sc-518218 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-518218 PE), fluorescein (sc-518218 FITC), Alexa Fluor 488 (sc-518218 AF488), Alexa Fluor 546 (sc-518218 AF546), Alexa Fluor 594 (sc-518218 AF594) or Alexa Fluor 647 (sc-518218 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor 680 (sc-518218 AF680) or Alexa Fluor 790 (sc-518218 AF790), 200 μ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

PDSS1 (C-8) is recommended for detection of PDSS1 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 PDSS1 siRNA (h): sc-62769, PDSS1 shRNA Plasmid (h): sc-62769-SH and PDSS1 shRNA (h) Lentiviral Particles: sc-62769-V.

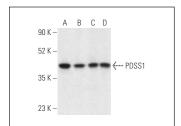
Molecular Weight of PDSS1: 46 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, HeLa whole cell lysate: sc-2200 or K-562 whole cell lysate: sc-2203.

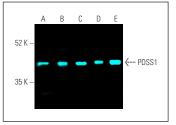
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-lgG κ BP-HRP: sc-516102 or m-lgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz MarkerTM 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-lgG κ BP-FITC: sc-516140 or m-lgG κ 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



PDSS1 (C-8): sc-518218. Western blot analysis of PDSS1 expression in Hep G2 (A), HeLa (B), K-562 (C) and Jurkat ($\dot{\rm D}$) whole cell lysates. Detection reagent used: m-lgG $_{\rm 2a}$ BP-HRP: sc-542731.



PDSS1 (C-8): sc-518218. Fluorescent western blot analysis of PDSS1 expression in HeLa (**A**), Jurkat (**B**), Hep G2 (**C**), K-562 (**D**) and HEK293T (**E**) whole cell lysates. Blocked with UltraCruz® Blocking Reagent: sc-516214. Detection reagent used: m-IgG_{2a} BP-CFL 647: sc-647738

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.