

Rabbit Anti-PDSS2 antibody

SL10178R

Product Name PDSS2

Chinese Name 抑癌蛋白 DLP1

Alias Il-trans-decaprenyl-diphosphate synthase subunit 2; bA59I9.3; C6orf210; Candidate tumor suppressor protein; chromosome 6 open reading frame 210; Decaprenyl pyrophosphate synthase subunit 2; decaprenyl pyrophosphate synthetase subunit 2; Decaprenyl-diphosphate synthase subunit 2; DLP1; DLP1_HUMAN; hDLP1; Pdss2; prenyl (decaprenyl) diphosphate synthase, subunit 2; subunit 2 of decaprenyl diphosphate synthase.

Research Area Tumour immunology Signal transduction Cell type markers Mitochondrion

Immunogen Species Rabbit

Clonality Polyclonal

React Species (predicted:Dog,Pig,Horse,Rabbit)
IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
(Paraffin sections need antigen repair)

Applications not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

Theoretical molecular weight 44kDa

Cellular localization cytoplasmic Mitochondrion

Form Liquid

Concentration 1mg/ml

immunogen KLH conjugated synthetic peptide derived from dog PDSS2: 31-130/399

Lsotype IgG

Purification affinity purified by Protein A

Buffer Solution 1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol.

Storage Shipped at 4°C. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.

Attention This product as supplied is intended for research use only, not for use in human, therapeutic or

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diagnostic applications.

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The protein encoded by this gene is an enzyme that synthesizes the prenyl side-chain of coenzyme Q, or ubiquinone, one of the key elements in the respiratory chain. The gene product catalyzes the formation of all trans-polyprenyl pyrophosphates from isopentyl diphosphate in the assembly of polyisoprenoid side chains, the first step in coenzyme Q biosynthesis. Defects in this gene are a cause of coenzyme Q10 deficiency.

Function:

Supplies decaprenyl diphosphate, the precursor for the side chain of the isoprenoid quinones ubiquinone-10.

Subunit:

Heterotetramer of 2 DPS1/TPRT and 2 DLP1 subunits.

Subcellular Location:

Mitochondrion (Potential).

DISEASE:

Defects in PDSS2 are the cause of coenzyme Q10 deficiency, primary, type 3 (COQ10D3) [MIM:614652]. A fatal encephalomyopathic form of coenzyme Q10 deficiency with nephritic syndrome. Coenzyme Q10 deficiency is an autosomal recessive disorder with variable manifestations consistent with 5 major phenotypes. The phenotypes include an encephalomyopathic form with seizures and ataxia; a multisystem infantile form with encephalopathy, cardiomyopathy and renal failure; a predominantly cerebellar form with ataxia and cerebellar atrophy; Leigh syndrome with growth retardation; and an isolated myopathic form.

Product Detail

Similarity:

Belongs to the FPP/GGPP synthase family.

SWISS:

Q07001

Gene ID:

481950

Database links:

[Entrez Gene: 57107](#) Human

[Entrez Gene: 71365](#) Mouse

[Entrez Gene: 365592](#) Rat



[Omim: 610564](#) Human

[SwissProt: Q86YH6](#) Human

[SwissProt: Q33DR3](#) Mouse

[SwissProt: Q5U2R1](#) Rat

[Unigene: 486095](#) Human

[Unigene: 363225](#) Mouse

[Unigene: 20063](#) Rat