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Rabbit Anti-PDSS2 antibody

SL10178R

Product Name:	PDSS2
Chinese Name:	抑癌蛋白DLP1
Alias:	ll-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.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Dog,Pig,Horse,Rabbit,
Applications:	ELISA=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:	44kDa
Cellular localization:	cytoplasmicMitochondrion
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from dog PDSS2:31-130/399
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:	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.

Similarity:
Belongs to the FPP/GGPP synthase family.

SWISS:
Q07001

Gene ID:
481950

Database links:

Entrez Gene: 57107Human

Entrez Gene: 71365Mouse

Entrez Gene: 365592Rat

Omim: 610564Human

SwissProt: Q86YH6Human

SwissProt: Q33DR3Mouse

SwissProt: Q5U2R1Rat

	Unigene: 486095Human Unigene: 363225Mouse Unigene: 20063Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.