10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

PDSS2 Rabbit pAb

Catalog # AP98534

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q86YH6
Other Accession	Q86YH6
Reactivity	Human
Predicted	Rat, Mouse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	44129
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human PDSS2
Epitope Specificity	21-100/399
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the FPP/GGPP synthase family.
SUBUNIT	Heterotetramer of 2 DPS1/TPRT and 2 DLP1 subunits.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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.

Additional Information

Gene ID	57107
Other Names	C6orf210; COQ10D3; COQ1B; DLP1; bA59I9.3; hDLP1; 5430420P03Rik; Gm60; Plmp; kd; mDLP1; DLP1_HUMAN; PDSS2; All-trans-decaprenyl-diphosphate synthase subunit 2; Candidate tumor suppressor protein; Decaprenyl pyrophosphate synthase subunit 2; Decaprenyl-diphosphate synthase subunit 2; Solanesyl-diphosphate synthase subunit 2; DLP1_MOUSE; DLP1_RAT;

	decaprenyl diphosphate synthase subunit 2; chromosome 6 open reading frame 210; prenyl (decaprenyl) diphosphate synthase, subunit 2
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	PDSS2 (HGNC:23041)
Synonyms	C6orf210, DLP1
Function	Heterotetrameric enzyme that catalyzes the condensation of farnesyl diphosphate (FPP), which acts as a primer, and isopentenyl diphosphate (IPP) to produce prenyl diphosphates of varying chain lengths and participates in the determination of the side chain of ubiquinone (PubMed:16262699). Supplies nona and decaprenyl diphosphate, the precursors for the side chain of the isoprenoid quinones ubiquinone-9 (Q9) and ubiquinone-10 (Q10) respectively (PubMed:16262699). The enzyme adds isopentenyl diphosphate molecules sequentially to farnesyl diphosphate with trans stereochemistry (PubMed:16262699). May play a role during cerebellar development (By similarity). May regulate mitochondrial respiratory chain function (By similarity).
Cellular Location	Mitochondrion.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.