抑癌蛋白 DLP1 抗体

产品货号： mlR0376

英文名称： PDSS2

中文名称： 抑癌蛋白 DLP1 抗体

别　　名： All-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.

研究领域： 肿瘤　免疫学　信号转导　细胞类型标志物　线粒体

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分 子 量： 44kDa

细胞定位： 细胞浆 线粒体

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human PDSS2:21-100/399

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： 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

产品介绍 background:

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:

Q86YH6

Gene ID:

57107

Important Note:

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

产品图片 ：