NDUFAF6 蛋白抗体

产品货号： mlR19077

英文名称： NDUFAF6

中文名称： NDUFAF6 蛋白抗体

别　　名： CCDC113; C8orf38; NDUF6_HUMAN; Coiled-coil domain-containing protein 113 Gene names; DKFZp434N1418; HSPC065; NADH dehydrogenase (ubiquinone) complex I assembly factor 6; Putative phytoene synthase; UPF0551 protein C8orf38 mitochondrial.

研究领域： 肿瘤　细胞生物　神经生物学　信号转导

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 ICC=1:100-500 IF=1:100-500 （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： 33kDa

细胞定位： 细胞核 细胞浆

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human NDUFAF6:201-300/333

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： This gene encodes a protein that localizes to mitochondria and contains a predicted phytoene synthase domain. The encoded protein plays an important role in the assembly of complex I (NADH-ubiquinone oxidoreductase) of the mitochondrial respiratory chain through regulation of subunit ND1 biogenesis. Mutations in this gene are associated with complex I enzymatic deficiency. [provided by RefSeq, Nov 2011]

Function:

Involved in the assembly of mitochondrial NADH:ubiquinone oxidoreductase complex (complex I) at early stages. May play a role in the biogenesis of MT-ND1.

Subcellular Location:

Isoform 1: Mitochondrion inner membrane. Note: Peripherally localized on the matrix face of the mitochondrial inner membrane. Ref.7

Isoform 2: Cytoplasm. Nucleus

DISEASE:

Mitochondrial complex I deficiency (MT-C1D) [MIM:252010]: A disorder of the mitochondrial respiratory chain

that causes a wide range of clinical manifestations from lethal neonatal disease to adult-onset neurodegenerative disorders. Phenotypes include macrocephaly with progressive leukodystrophy, non-specific encephalopathy, cardiomyopathy, myopathy, liver disease, Leigh syndrome, Leber hereditary optic neuropathy, and some forms of Parkinson disease. Note: The disease is caused by mutations affecting the gene represented in this entry.

Similarity:

Belongs to the NDUFAF6 family.

SWISS:

Q330K2

Gene ID:

137682

Important Note:

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