细胞色素氧化酶缺失蛋白 2 抗体

产品货号： mlR17295

英文名称： SCO2

中文名称： 细胞色素氧化酶缺失蛋白 2 抗体

别　　名： Cytochrome oxidase deficient homolog 2; MGC125823; MGC125825; OTTHUMP00000196774; OTTHUMP00000196775; Protein SCO2 homolog, mitochondrial; SCO (cytochrome oxidase deficient, yeast) homolog 2; SCO 1L; SCO 2; SCO cytochrome oxidase deficient homolog 2 (yeast); SCO cytochrome oxidase deficient homolog 2; SCO1L; SCO2; SCO2_HUMAN; Synthesis of cytochrome c oxidase 2.

研究领域： 肿瘤　细胞生物　信号转导　新陈代谢　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 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 ： 25kDa

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 SCO2:201-266/266

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 : Cytochrome c oxidase (COX) catalyzes the transfer of electrons from cytochrome c to molecular oxygen, which helps to maintain the proton gradient across the inner mitochondrial membrane that is necessary for aerobic ATP production. Human COX is a multimeric protein complex that requires several assembly factors; this gene encodes one of the COX asembly factors. The encoded protein is a metallochaperone that is involved in the biogenesis of cytochrome c oxidase subunit II. Mutations in this gene are associated with fatal infantile encephalocardiomyopathy. [provided by RefSeq, Dec 2009]

Function:

Acts as a copper chaperone, transporting copper to the Cu(A) site on the cytochrome c oxidase subunit II (COX2).

Subcellular Location:

Mitochondrion.

Tissue Specificity:

Ubiquitous.

DISEASE:

Defects in SCO2 are the cause of fatal infantile cardioencephalomyopathy with cytochrome c oxidase deficiency (FIC) [MIM:604377]. This disease is characterized by hypertrophic cardiomyopathy, lactic acidosis, and gliosis. Heart and skeletal muscle show reductions in cytochrome c oxidase (COX) activity, whereas liver and fibroblasts show mild COX deficiencies.

Similarity:

Belongs to the SCO1/2 family.

Contains 1 thioredoxin domain.

SWISS:

O43819

Gene ID:

9997

Important Note:

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