琥珀酸脱氢酶复合体亚基 A 抗体

产品货号： mlR3970

英文名称： SDHA

中文名称： 琥珀酸脱氢酶复合体亚基 A 抗体

别　　名： mitochondrial; DHSA_HUMAN; Flavoprotein subunit of complex II; Fp; SDH 2; SDH2; SDHA; SDHF; Succinate dehydrogenase [ubiquinone] flavoprotein subunit; Succinate dehydrogenase [ubiquinone] flavoprotein subunit mitochondrial; Succinate dehydrogenase complex flavoprotein subunit precursor; Succinate dehydrogenase complex subunit A; Succinate Dehydrogenase Complex subunit A Flavoprotein.

研究领域： 肿瘤　细胞生物　免疫学　信号转导　细胞凋亡　转录调节因子

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat,

产品应用： WB=1:500-2000 ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 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： 70kDa

细胞定位： 细胞浆

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human SDHA:561-664/664

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:

In aerobic respiration reactions, succinate dehydrogenase (SDH) catalyzes the oxidation of succinate and ubiquinone to fumarate and ubiquinol. Four subunits comprise the SDH protein complex: a flavochrome subunit (SDHA), an iron-sulfur protein (SDHB), and two membrane-bound subunits (SDHC and SDHD) anchored to the inner mitochondrial membrane. Mutations to these subunits cause mitochondrial dysfunction, corresponding to several distinct disorders. Mutations in the membrane bound components may cause hereditary paraganglioma, while SDHA mutations are associated with juvenile encephalopathy as well as Leigh Syndrome, a severe neurological disorder. Inactivating mutations in SDHB correlate with inherited, but not necessarily sporadic, cases of pheochromocytoma.

Function:

Defects in SDHA are a cause of mitochondrial complex II deficiency (MT-C2D). A disorder of the mitochondrial respiratory chain with heterogeneous clinical manifestations. Clinical features include psychomotor regression in infants, poor growth with lack of speech development, severe spastic quadriplegia, dystonia, progressive leukoencephalopathy, muscle weakness, exercise intolerance, cardiomyopathy. Some patients manifest Leigh syndrome or Kearns-Sayre syndrome.

SWISS:

P31040

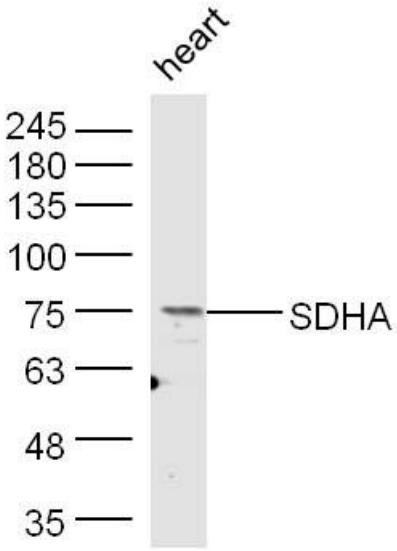
Gene ID:

6389

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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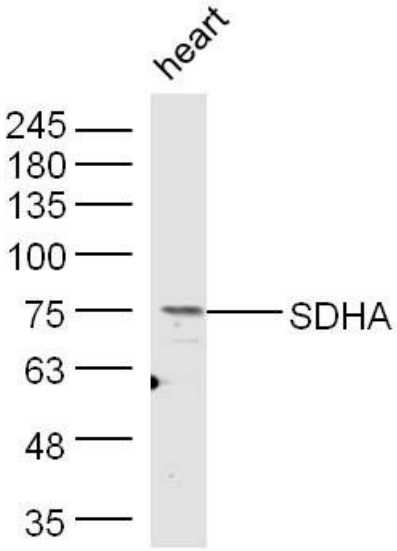
180
135
100
75
63
48
35
SDHA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