视神经萎缩相关蛋白 1 抗体

产品货号： mlR11764

英文名称： OPA1

中文名称： 视神经萎缩相关蛋白 1 抗体

别　　名： Dynamin like 120 kDa protein; Dynamin like 120 kDa protein, mitochondrial; Dynamin-like 120 kDa protein; Dynamin-like 120 kDa protein, form S1; FLJ12460; Juvenile kjer type optic atrophy; Juvenile kjer-type optic atrophy; KIAA0567; KJER type; Large GTP binding protein; largeG; MGM1; Mitochondrial dynamin like 120 kDa protein; Mitochondrial dynamin like GTPase; NPG; NTG; OAK; OPA 1; OPA1; OPA1 gene; OPA1_HUMAN; Optic atrophy 1 (autosomal dominant); OPTIC ATROPHY 1; Optic atrophy 1 gene protein; Optic atrophy 1 homolog (human); Optic atrophy protein 1; Optic atrophy protein 1 homolog.

研究领域： 心血管　细胞生物　神经生物学

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Dog, Pig, Cow, Horse, Rabbit, Sheep,

产品应用： WB=1:500-2000 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： 111kDa

细胞定位： 细胞浆 细胞膜

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human OPA1:651-750/960

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： OPA1 is a 120kDa protein belonging to the dynamin family. The OPA1 gene has been localized to 3q29. The gene is targeted to mitochondria and is involved in mitochondrial biogenesis. Defects in OPA1 are a cause of optic atrophy type 1. OPA1 is mostly expressed in retina but can also be expressed in brain, testis, heart and skeletal muscle.

Function:

Dynamin-related GTPase required for mitochondrial fusion and regulation of apoptosis. May form a diffusion barrier for proteins stored in mitochondrial cristae. Proteolytic processing in response to intrinsic apoptotic signals may lead to disassembly of OPA1 oligomers and release of the caspase activator cytochrome C (CYCS) into the mitochondrial intermembrane space.

Subcellular Location:

Mitochondrion inner membrane. Mitochondrion intermembrane space.

Tissue Specificity:

Highly expressed in retina. Also expressed in brain, testis, heart and skeletal muscle. Isoform 1 expressed in retina, skeletal muscle, heart, lung, ovary, colon, thyroid gland, leukocytes and fetal brain. Isoform 2 expressed in colon, liver, kidney, thyroid gland and leukocytes. Low levels of all isoforms expressed in a variety of tissues.

Post-translational modifications:

PARL-dependent proteolytic processing releases an antiapoptotic soluble form not required for mitochondrial fusion.

DISEASE:

Defects in OPA1 are a cause of optic atrophy type 1 (OPA1) [MIM:165500]. OPA1 is a dominantly inherited optic neuropathy occurring in 1 in 50,000 individuals that features progressive loss in visual acuity leading, in many cases, to legal blindness.

Defects in OPA1 are the cause of optic atrophy 1 with deafness (OPA1D) [MIM:125250]. Some individuals with mutations in OPA1 manifest also ophthalmoplegia and myopathy.

Similarity:

Belongs to the dynamin family.

SWISS:

O60313

Gene ID:

4976

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