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Rabbit Anti-OPA1/AF350 Conjugated antibody

SL11764R-AF350

Product Name	Anti-OPA1/AF350
Chinese Name	AF350 标记的视神经萎缩相关蛋白 1 抗体
Alias	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.
Research Area	Cardiovascular Cell biology Neurobiology
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	Rat(predicted:Human,Mouse,Dog,Pig,Cow,Horse,Rabbit,Sheep)
Applications	IF=1:100-500 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight	111kDa
Form	Lyophilized or Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human OPA1
Lsotype	IgG
Purification	affinity purified by Protein A
Storage Buffer	1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol
Storage	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 1M PBS or diluent of antibody the antibody is stable for at least two weeks

at 2-4 °C.

Product Detail

background:
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.

Database links:

Entrez Gene: 424900 Chicken

Entrez Gene: 4976 Human

Entrez Gene: 74143 Mouse

Entrez Gene: 171116 Rat

Omim: 605290 Human

SwissProt: O60313 Human

SwissProt: P58281 Mouse

SwissProt: Q2TA68 Rat

Unigene: 594504 Human

Unigene: 274285 Mouse

Unigene: 9783 Rat

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