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SCO1 Rabbit pAb

Catalog # AP99809

Product Information

Application	IHC-P, IHC-F, IF, E
Primary Accession	O75880
Other Accession	O75880
Predicted	Rat, Human, Mouse, Rabbit, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	33814
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human SCO1
Epitope Specificity	201-301/301
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the SCO1/2 family.
DISEASE	Defects in SCO1 are a cause of mitochondrial complex IV deficiency (MT-C4D) [MIM:220110]; also known as cytochrome c oxidase deficiency. A disorder of the mitochondrial respiratory chain with heterogeneous clinical manifestations, ranging from isolated myopathy to severe multisystem disease affecting several tissues and organs. Features include hypertrophic cardiomyopathy, hepatomegaly and liver dysfunction, hypotonia, muscle weakness, excercise intolerance, developmental delay, delayed motor development and mental retardation. A subset of patients manifest Leigh syndrome.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Mammalian cytochrome c oxidase (COX) catalyzes the transfer of reducing equivalents from cytochrome c to molecular oxygen and pumps protons across the inner mitochondrial membrane. In yeast, 2 related COX assembly genes, SCO1 and SCO2 (synthesis of cytochrome c oxidase), enable subunits 1 and 2 to be incorporated into the holoprotein. This gene is the human homolog to the yeast SCO1 gene. [provided by RefSeq, Jul 2008]

Additional Information

Gene ID	6341
Other Names	MC4DN4; SCOD1; 2610001C07Rik; D11Bwg1310e; RGD1559538; SCO1_HUMAN; SCO1; SCO1_MOUSE; synthesis of cytochrome C oxidase 1; SCO (cytochrome oxidase deficient, yeast) homolog 1; SCO cytochrome oxidase deficient homolog 1 (yeast); SCO1, cytochrome c oxidase assembly protein; SCO cytochrome c oxidase assembly protein 1
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:500

	0-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	SCO1
Synonyms	SCOD1
Function	Copper metallochaperone essential for the maturation of cytochrome c oxidase subunit II (MT-CO2/COX2). Together with SCO2, involved in delivering copper to the Cu(A) site on MT-CO2/COX2 (PubMed:15229189, PubMed:15659396, PubMed:16735468, PubMed:17189203, PubMed:19336478). Plays an important role in the regulation of copper homeostasis by controlling the abundance and cell membrane localization of copper transporter CTR1 (By similarity).
Cellular Location	Mitochondrion inner membrane; Single-pass membrane protein
Tissue Location	Predominantly expressed in tissues characterized by high rates of oxidative phosphorylation (OxPhos), including muscle, heart, and brain.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.