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Rabbit Anti-CPT2/Cy5 Conjugated antibody

SL5047R-Cy5

Product Name	Anti-CPT2/Cy5
Chinese Name	Cy5 标记的肉毒碱棕榈酰基转移酶 2 抗体
Alias	Carnitine O palmitoyltransferase 2; Carnitine O palmitoyltransferase 2 mitochondrial; Carnitine O-palmitoyltransferase 2; Carnitine palmitoyltransferase II; CPT 1; CPT 2; CPT II; CPT1; CPT2; CPT2_HUMAN; CPTASE; CPTII; mitochondrial.
Research Area	Cardiovascular Cell biology immunology Signal transduction Cell type markers Lipoprotein The new supersedes the old Mitochondrion
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	(predicted:Human,Mouse,Rat,Dog,Pig,Horse,Rabbit)
Applications	IF=1:100-500 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight	71kDa
Form	Lyophilized or Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human CPT2
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:** The protein encoded by this gene is a nuclear protein which is transported to the mitochondrial inner membrane. Together with carnitine palmitoyltransferase I, the encoded protein oxidizes long-chain fatty acids in the mitochondria. Defects in this gene are associated with mitochondrial

long-chain fatty-acid (LCFA) oxidation disorders. [provided by RefSeq, Jul 2008].

Subcellular Location:
Mitochondrion inner membrane; Peripheral membrane protein; Matrix side.

DISEASE:
Carnitine palmitoyltransferase 2 deficiency late-onset (CPT2D) [MIM:255110]: Autosomal recessive disorder characterized by recurrent myoglobinuria, episodes of muscle pain, stiffness, and rhabdomyolysis. These symptoms are triggered by prolonged exercise, fasting or viral infection and patients are usually young adults. In addition to this classical, late-onset, muscular type, a hepatic or hepatocardiomuscular form has been reported in infants. Clinical pictures in these children or neonates include hypoketotic hypoglycemia, liver dysfunction, cardiomyopathy and sudden death. Note=The disease is caused by mutations affecting the gene represented in this entry.
Carnitine palmitoyltransferase 2 deficiency infantile (CPT2DI) [MIM:600649]: A disorder of mitochondrial long-chain fatty acid oxidation characterized by hepatic or hepato-cardio-muscular manifestations with onset in infancy. Clinical features include hypoketotic hypoglycemia, lethargy, seizures, hepatomegaly, liver dysfunction, cardiomegaly and dilated cardiomyopathy. Note=The disease is caused by mutations affecting the gene represented in this entry.
Carnitine palmitoyltransferase 2 deficiency lethal neonatal (CPT2D-LN) [MIM:608836]: Lethal neonatal form of CPT2D. This rarely presentation is antenatal with cerebral periventricular cysts and cystic dysplastic kidneys. The clinical variability of the disease is likely attributed to the variable residual enzymatic activity. Note=The disease is caused by mutations affecting the gene represented in this entry.
Encephalopathy, acute, infection-induced, 4 (IIAE4) [MIM:614212]: A severe neurologic complication of an infection. It manifests within days in otherwise healthy children after common viral infections, without evidence of viral infection of the brain or inflammatory cell infiltration. In affected children, high-grade fever is accompanied within 12 to 48 hours by febrile convulsions, often leading to coma, multiple-organ failure, brain edema, and high morbidity and mortality. The infections are usually viral, particularly influenza, although other viruses and even mycoplasma have been found to cause the disorder. Note=Disease susceptibility is associated with variations affecting the gene represented in this entry. CPT2 polymorphic variants do not cause classical carnitine palmitoyltransferase 2 deficiency, and patients harboring any of them are asymptomatic most of the time. However, they are prone to viral infection (high fever)-related encephalopathy (PubMed:21697855).

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Similarity:
Belongs to the carnitine/choline acetyltransferase family.

Database links:
UniProtKB/Swiss-Prot: P23786.2

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