

SPTLC2 Rabbit pAb

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Product Information

Application WB, IHC-P, IHC-F, IF, E

Primary Accession <u>015270</u>

Reactivity Rat, Pig, Mouse, Dog

Host Rabbit
Clonality Polyclonal
Calculated MW 62924
Physical State Liquid

Immunogen KLH conjugated synthetic peptide derived from human SPTLC2

Epitope Specificity 301-400/562

Isotype IgG

Purity affinity purified by Protein A

Buffer SUBCELLULAR LOCATION

SUBCELLULAR LOCATION

SIMILARITY

0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. Endoplasmic reticulum membrane; Single-pass membrane protein. Belongs to the class-II pyridoxal-phosphate-dependent aminotransferase

family.

SUBUNIT eterodimer with SPTLC1. Component of the serine palmitoyltransferase (SPT)

complex, composed of LCB1/SPTLC1, LCB2 (SPTLC2 or SPTLC3) and ssPT

(C14orf147/SSSPTA and C3orf57/SSSPTB).

DISEASE Defects in SPTLC2 are the cause of hereditary sensory and autonomic

neuropathy type 1C (HSAN1C) [MIM:613640]. It is a form of hereditary

sensory and autonomic neuropathy, a genetically and clinically

heterogeneous group of disorders characterized by degeneration of dorsal root and autonomic ganglion cells, and by prominent sensory abnormalities with a variable degree of motor and autonomic dysfunction. The neurological phenotype is often complicated by severe infections, osteomyelitis, and amputations. HSAN1C symptoms include loss of touch and vibration in the feet, dysesthesia and severe panmodal sensory loss in the upper and lower limbs, distal lower limb sensory loss with ulceration and osteomyelitis, and

distal muscle weakness.

Important Note This product as supplied is intended for research use only, not for use in

human, therapeutic or diagnostic applications.

Background Descriptions This gene encodes a long chain base subunit of serine palmitoyltransferase.

Serine palmitoyltransferase, which consists of two different subunits, is the

key enzyme in sphingolipid biosynthesis. It catalyzes the

pyridoxal-5-prime-phosphate-dependent condensation of L-serine and palmitoyl-CoA to 3-oxosphinganine. Mutations in this gene were identified in patients with hereditary sensory neuropathy type I. [provided by RefSeq, Mar

2011].

Additional Information

Gene ID 9517

Other Names Serine palmitoyltransferase 2, 2.3.1.50, Long chain base biosynthesis protein

2, LCB 2, Long chain base biosynthesis protein 2a, LCB2a,

Serine-palmitoyl-CoA transferase 2, SPT 2, SPTLC2 (HGNC:11278), KIAA0526,

LCB2

Target/Specificity Widely expressed.

Dilution WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,ELISA=1:5000

-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 SPTLC2 (HGNC:11278)

Synonyms KIAA0526, LCB2

Function Component of the serine palmitoyltransferase multisubunit enzyme (SPT)

that catalyzes the initial and rate-limiting step in sphingolipid biosynthesis by condensing L-serine and activated acyl-CoA (most commonly palmitoyl-CoA)

to form long-chain bases (PubMed: 19416851, PubMed: 19648650,

PubMed: 20504773, PubMed: 20920666). The SPT complex is composed of SPTLC1, SPTLC2 or SPTLC3 and SPTSSA or SPTSSB. Within this complex, the heterodimer consisting of SPTLC1 and SPTLC2/SPTLC3 forms the catalytic core (PubMed: 19416851). The composition of the serine palmitoyltransferase (SPT) complex determines the substrate preference (PubMed: 19416851). The SPTLC1-SPTLC2-SPTSSA complex shows a strong preference for C16-CoA substrate, while the SPTLC1-SPTLC3-SPTSSA isozyme uses both C14-CoA and

C16-CoA as substrates, with a slight preference for C14-CoA

(PubMed:<u>19416851</u>, PubMed:<u>19648650</u>). The SPTLC1-SPTLC2-SPTSSB complex

shows a strong preference for C18-CoA substrate, while the

SPTLC1-SPTLC3-SPTSSB isozyme displays an ability to use a broader range of

acyl-CoAs, without apparent preference (PubMed: <u>19416851</u>, PubMed: <u>19648650</u>). Crucial for adipogenesis (By similarity).

Cellular Location Endoplasmic reticulum membrane {ECO:0000250 | UniProtKB:P97363};

Single-pass membrane protein {ECO:0000250 | UniProtKB:P97363}

Tissue Location Widely expressed..

Background

This gene encodes a long chain base subunit of serine palmitoyltransferase. Serine palmitoyltransferase, which consists of two different subunits, is the key enzyme in sphingolipid biosynthesis. It catalyzes the pyridoxal-5-prime-phosphate-dependent condensation of L-serine and palmitoyl-CoA to 3-oxosphinganine. Mutations in this gene were identified in patients with hereditary sensory neuropathy type I. [provided by RefSeq, Mar 2011].

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