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# Anti-SPTLC3 Antibody from rabbit

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| Art. ID | SAF-ABS1640 |
| Unit | EA |

Description

Serine palmitoyltransferase 3 (EC 2.3.1.50, UniProt Q9NUV7, also known as LCB 3, LCB2b, Long chain base biosynthesis protein 2b, Long chain base biosynthesis protein 3, Serine-palmitoyl-CoA transferase 3, SPT 3) is encoded by the SPTLC3 (also known as C20orf38, SPTLC2L) gene (Gene ID 55304) in human. Serine palmitoyltransferase (SPT) is a pyridoxal 5'-phosphate (PLP)-dependent enzyme that catalyses the first step of de novo sphingolipids (SLs) biosynthesis, namely the condensation of L-serine with palmitoyl coenzyme-A to generate 3-ketodehydrosphinganine (KDS) that forms the sphingoid base backbone of all SLs. The core human SPT homoenzyme is a membrane-bound heterodimer composed of an hLCB1 (SPTLC1) subunit together with either an hLCB2a (SPTLC2) or an hLCB2b (SPTLC3) subunit. The hLCB1/hLCB2 heterodimer contains a single active site composed of residues from both subunits. hLCB2a and hLCB2b share 68% sequence identity, with hLCB2b being the isotype that is predominantly expressed in the placenta and prefers shorter chain acyl-CoAs (C12 and C14) to generate short-chain SLs. In addition, two human small subunits (ssSPTa and ssSPTb) have been identified and found to maximize the catalytic activity and to confer the acyl-CoA substrate chain-length specificity of the hLCB1/hLCB2 core enzyme. Mutations in hLCB1 and hLCB2a are found in patients with hereditary sensory and autonomic neuropathy type I (HSAN1), an inherited disorder that affects sensory and autonomic neurons due to neurotoxic deoxysphingolipids formation as a result of substrate promiscuity of the mutated SPT enzyme.