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Deoxysphingolipid

National Cancer Institute

Source

National Cancer Institute. *Deoxysphingolipid*. NCI Thesaurus. Code C116391.

An atypical, cytotoxic sphingolipid produced during abnormal de novo sphingolipid synthesis in which the enzyme serine palmitoyltransferase (SPT) is mutated. During de novo sphingolipid synthesis, SPT normally converts L-serine and palmitoyl-Coenzyme A into 3-keto-dihydrosphingosine, which is processed further to form ceramide. Mutated forms of SPT can accommodate the alternative substrates L-alanine or L-glycine, which leads to the formation of the deoxysphingolipids, deoxysphinganine and deoxymethylsphinganine, respectively. These atypical lipids are neither converted to ceramide nor degraded by the cellular glycosphingolipid hydrolysis pathways. Therefore, deoxysphingolipids can accumulate to neurotoxic levels, which can lead to neuropathy. Elevated plasma levels of deoxysphingolipids may be associated with diabetes or metabolic syndrome.