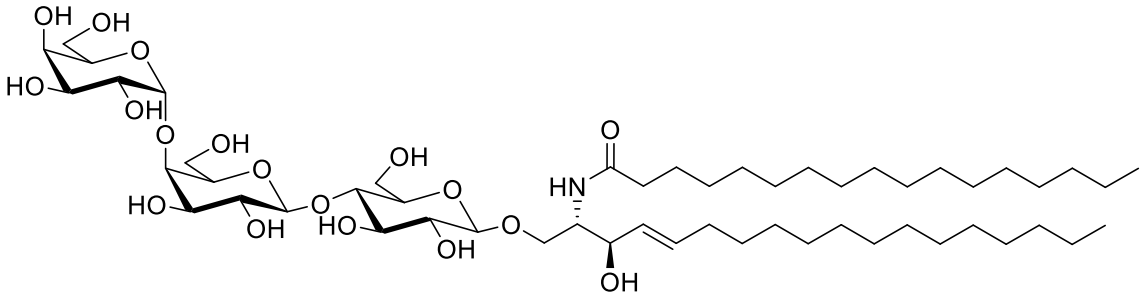
PRODUCT DATA SHEET

N-Heptadecanoyl-ceramide trihexoside

Catalog number: 1523
Synonyms: N-C17:0-Ceramide trihexoside; N-Heptadecanoyl globotriaosylceramide
Source: semisynthetic, porcine RBCs
Solubility: chloroform/methanol 2:1; DMSO; hot methanol
CAS number: 536745-81-0

Molecular Formula: $C_{53}H_{99}NO_{18}$
Molecular Weight: 1038
Storage: -20˚C
Purity: TLC: >98%, HPLC: >98%; identity confirmed by MS
TLC System: chloroform/methanol/DI water (65:25:4 by vol.)
Appearance: solid



Application Notes:

This product is a well-defined CTH containing the uncommon heptadecanoic acid acylated to the ceramide making it ideal as an internal standard and for biological studies.[1] Ceramide trihexoside (CTH) is a glycosphingolipid found mostly in mammalian cell membranes. It is involved in cellular signaling and has been identified as a receptor for various toxins including shiga toxins and shiga-like toxins.[2] Some toxins, such as veratoxins from *Escherichia coli*, require specific fatty acids on the ceramide portion of CTH to show affinity in binding. An accumulation of CTH in the cellular membranes due to a lack of *alpha*-galactosidase to convert it into lactosyl ceramide results in Fabry disease.[3] This product can be used as an excellent standard for the identification of CTH in Fabry disease by HPLC[4] and mass spectrometry. An inability to convert CTH to globoside due to mutations in the gene sequence leads to the P^k blood group phenotype. It appears that under certain conditions CTH can enhance anticoagulant activity. CTH has also been studied as a tool to investigate lymphocyte activation.[5]

Selected References:

1. M. Fuller et al. "Urinary Lipid Profiling for the Identification of Fabry Hemizygotes and Heterozygotes" *Clinical Chemistry*, Vol. 51 pp. 688-694, *2005*
2. S. Ashkenazi and T. G. Cleary, "Rapid method to detect shiga toxin and shiga-like toxin I based on binding to globotriosyl ceramide (Gb3), their natural receptor." *J Clin Microbio*. June; 27(6): 1145-1150, *1989*
3. S. Bekri, O. Lidove, R. Jaussaud, B. Knebelmann, F. Barbey. "The role of ceramide trihexoside (globotriaosylceramide) in the diagnosis and follow-up of the efficacy of treatment of Fabry disease: a review of the literature". *Cardiovasc Hematol Agents Med Chem* 4 (4): 289–97, *October 2006*
4. J. E. Groener, B. J. Poorthuis, S. Kuiper, M. T. Helmond, C. E. Hollak, J. M. Aerts. "HPLC for simultaneous quantification of total ceramide, glucosylceramide, and ceramide trihexoside concentrations in plasma." *Clin Chem.*, Apr;53(4):742-7, *2007*. Epub Mar 1 *2007*
5. C. Menge, I. Stamm, M. Wuhrer, R. Geyer, L. H. Wieler, G. Baljer. "Globotriaosylceramide (Gb(3)/CD77) is synthesized and surface expressed by bovine lymphocytes upon activation in vitro." *Vet Immunol Immunopathol.*, Nov;83(1-2):19-36, *2001*

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