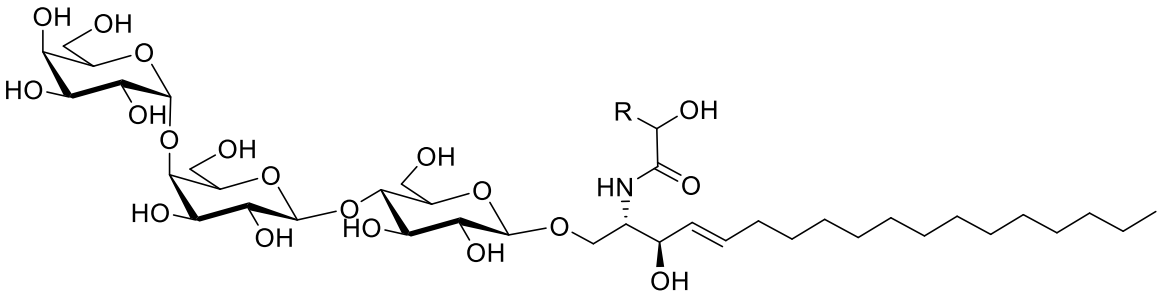
PRODUCT DATA SHEET

Ceramide trihexosides (bottom spot)

Catalog number: 1514
Common Name: CTH with hydroxy fatty acid side chain
Source: natural, porcine RBC
Solubility: hot methanol,DMSO, chloroform/methanol, 1:1
CAS number: N/A

Molecular Formula: $C_{60}H_{113}NO_{19}$
Molecular Weight: 1153 (hydroxytetracosanoyl)
Storage: -20°C
Purity: TLC >98%; identity confirmed by MS
TLC System: chloroform/methanol/DI water, (65:25:4 by vol.)
Appearance: solid



Application Notes:

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.[1] 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.[2] This product can be used as an excellent standard for the identification of CTH in Fabry disease by HPLC[3] and mass spectrometry.[4] 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] Neutral glycolipids may help to cause resistance to natural killer cells in some types of cells. Interferon appears to slightly increase the resistance of cells towards natural killer cells and also increases the amounts of non-hydroxylated fatty acids on various glycolipids, suggesting that there may be a connection between the hydroxylated fatty acids and natural killer cell resistance.

Selected References:

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2. 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*
3. 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*
4. K. Mills, A. Johnson, B. Winchester. "Synthesis of novel internal standards for the quantitative determination of plasma ceramide trihexoside in Fabry disease by tandem mass spectrometry." *FEBS Lett.*, Mar 27;515(1-3):171-6, *2002*
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*