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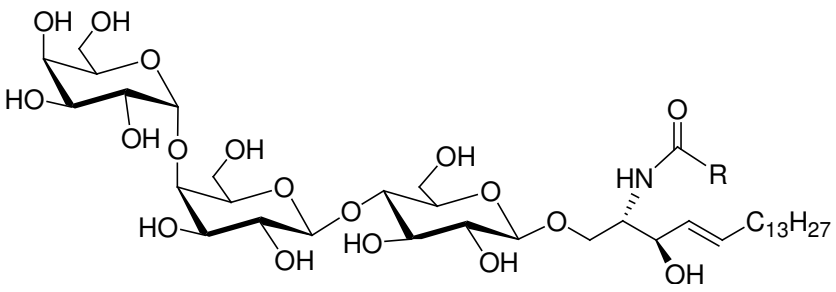
# PRODUCT DATA SHEET

## Ceramide trihexosides (top spot)

**Catalog No:** 1513
**Common Name:** CTH with non-hydroxy fatty acid side chain
**Source:** natural, porcine RBCs
**Solubility:** chloroform/methanol (2:1)
**CAS No:** N/A
**Molecular Formula:** $C_{54}H_{101}NO_{18}$

**Molecular Weight:** 1052 (stearoyl)
**Storage:** -20°C
**Purity:** TLC > 98%; identity confirmed by MS
**TLC System:** chlorform/methanol/DI water (65:25:4 by vol.)
**Appearance:** solid




### Application Notes:

Ceramide trihexoside 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]

### Selected References:

1. 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 Microbiol,* Vol. 27:6 pp. 1145-1150, *1989*
2. S. Bekri et al. "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,* Vol. 4:4 pp. 289–297, *2006*
3. J. Groener et al. "HPLC for simultaneous quantification of total ceramide, glucosylceramide, and ceramide trihexoside concentrations in plasma.". *Clin Chem.,* Vol. 53:4 pp.742-747, *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.,* Vol. 515 pp. 171-176, *2002*
5. C. Menge et al. "Globotriaosylceramide (Gb(3)/CD77) is synthesized and surface expressed by bovine lymphocytes upon activation in vitro." *Vet Immunol Immunopathol.,* Vol. 83 pp.19-36, *2001*