

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: C₅₄H₁₀₁NO₁₈

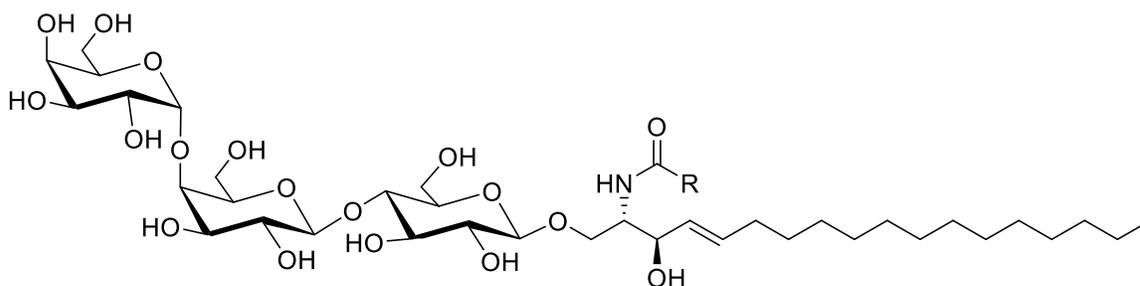
Molecular Weight: 1052 (stearoyl)

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 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 HPLC³ 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

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