## PRODUCT DATA SHEET



## N-Hexadecanoyl-D<sub>9</sub> (13,13,14,14,15,15,16,16,16)-ceramide trihexosides

Catalog number: 1551

**Synonyms:** CTH-D<sub>9</sub>; Gb<sub>3</sub>-D<sub>9</sub>;

Globotriaosylceramide-D<sub>9</sub>; N-C16:0-CD<sub>9</sub>-CTH; N-C16:0-CD<sub>9</sub>-Gb<sub>3</sub>; N-Hexadecanoyl-CD<sub>9</sub>-globotriaosylceramide; N-Palmitoyl-CD<sub>9</sub>-ceramide

trihexoside

**Source:** semisynthetic, porcine RBC

**Solubility:** Chloroform/methanol/water, 2:1:0.1;

DMSO

CAS number: N/A

Molecular Formula: C<sub>52</sub>H<sub>88</sub>D<sub>9</sub>NO<sub>18</sub>

**Molecular Weight:** 1033

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:**

This high purity deuterated product is ideal for the identification of ceramide trihexoside in samples and biological systems. 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. 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. This product can be used as an excellent standard for the identification of CTH in Fabry disease by HPLC<sup>3</sup> and mass spectrometry. An inability to convert CTH to globoside due to mutations in the gene sequence leads to the P<sup>k</sup> 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.

## **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
- 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

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