PRODUCT DATA SHEET



N-Tricosanoyl ceramide trihexoside

Catalog No: 1524

Common Name: N-C23:0-Ceramide

trihexoside; N-Tricosanoyl globotriaosylceramide

Source: semisynthetic, porcine RBC **Solubility:** chloroform/methanol (2:1)

DMSO, hot methanol

CAS No: 536745-84-3

Molecular Formula: C₅₉H₁₁₁NO₁₈

Molecular Weight: 1123

Storage: -20°C

Purity: TLC > 98%; identity confirmed by MS **TLC System:** chloroform/methanol/DI water

(65:25:3 by Vol.)

Appearance: solid

Application Notes:

This product is a well-defined ceramide trihexoside containing a tricosanoic fatty acid acyl group on the sphingosine. 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. 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 and mass spectrometry. An inability to convert CTH to globoside due to mutations in the gene sequence leads to the Pk 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:

- 1. S. Ashkenazi, and T. 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. 27 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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