

PRODUCT DATA SHEET

N-Glycinated *lyso*-ceramide trihexoside

Catalog number: 1530

Synonyms: N-Glycinated
globotriaosylsphingosine; Internal
standard for
globotriaosylsphingosine

Source: semisynthetic, porcine RBC

Solubility: chloroform/methanol/DI water
2:1:0.1, DMF

CAS number: N/A

Molecular Formula: C₃₈H₇₀N₂O₁₈

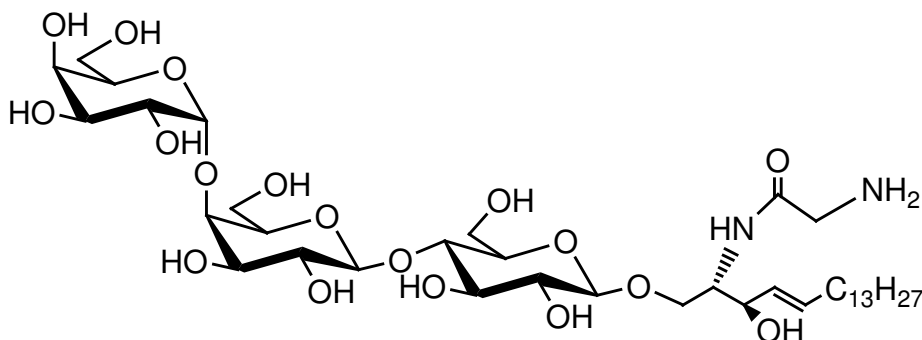
Molecular Weight: 843

Storage: -20°C

Purity: 98⁺%; identity confirmed by MS

TLC System: chloroform/methanol/DI
water/2.5 N ammonium
hydroxide, 60:40:7:3

Appearance: solid



Application Notes:

N-Glycinated *lyso*-ceramide trihexoside is an analogue of the important biomolecule *lyso*-ceramide trihexoside (globotriaosylsphingosine, *lyso*-Gb3). It is ideal for use as an internal standard in the extraction and mass spectrometry (MS) analysis of *lyso*-ceramide trihexoside.⁽¹⁾ The free amine group gives this product very similar physical characteristics to the natural *lyso*-ceramide trihexoside while the glycine adds an additional 57 units to the molecule making it easy to detect by MS. *lyso*-Ceramide trihexoside and the acylated ceramide trihexoside (globotriaosylceramide, Gb3) are important biomarkers for the lysosomal storage disorder Fabry disease.⁽²⁾ Fabry disease is characterized by a deficiency in the enzyme α -galactosidase, resulting in an accumulation of ceramide trihexoside and *lyso*-ceramide trihexoside.⁽³⁾ Early detection and treatment of this disease is critical to prevent damage to various organs.

Selected References:

1. R. Krüger et al. Quantification of the Fabry marker lysoGb3 in human plasma by tandem mass spectrometry. *Journal of Chromatography B.*, Vol. 883-884, pp. 128-135, 2012
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.*, Vol. 4:4 pp. 289-297, 2006
3. C. Auray-Blais et al. How well does urinary lyso-Gb3 function as a biomarker in Fabry disease. *Clin. Chim. Acta*, Vol. 411(23-24) pp. 1906-1914, 2010

This product is to be used for research only. It is not intended for drug or diagnostic use, human consumption or to be used in food or food additives. Matreya assumes no liability for any use of this product by the end user. We believe the information, offered in good faith, is accurate.