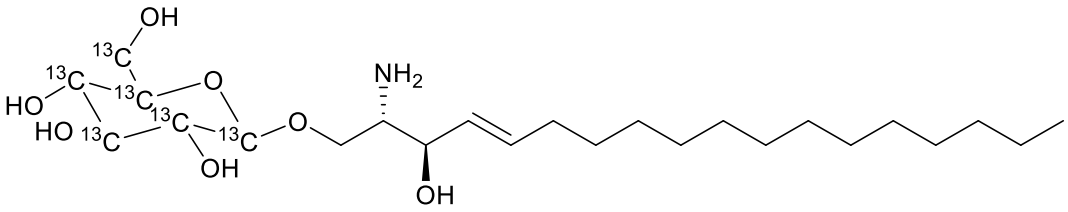
PRODUCT DATA SHEET

^{13}C6-Glucosylsphingosine

Catalog No: 2209
Common Name: 1-(*beta*-D-Glucosyl-1,2,3,4,5,6-$^{13}C_6$)-sphingosine; $^{13}C_6$-*lyso*-glucocerebroside
Source: synthetic
Solubility: ethanol, methanol, chloroform/methanol, 2:1
CAS No: 299172-48-8

Molecular Formula: $C_{18}{}^{13}C_6H_{47}NO_7$
Molecular Weight: 468
Storage: -20°C
Purity: TLC, HPLC > 98%; identity confirmed by MS
TLC System: chloroform/methanol/2.5N ammonium hydroxide (65:25:4 by vol.)
Appearance: solid



Application Notes:

Glucosylsphingosine is the *lyso*-derivative of the common glycolipid glucocerebroside. This product is a stable isotope labeled standard containing six carbon-13 units on the glucose moiety. Glucosylceramide and glucosylsphingosine are important biological species that are the precursors for many complex glycosphingolipids. These lipids are involved in critical cellular functions such as cellular proliferation, differentiation, adhesion, signal transduction, cell-to-cell interactions, tumorigenesis, and metastasis. One of the most important roles of glucosylsphingosine is as a biomarker for the lysosomal storage disorder Gaucher disease.

Gaucher disease is characterized by an accumulation of glucocerebroside due to a deficiency in the enzyme glucocerebrosidase and it has now been found that glucosylsphingosine also accumulates in this disease.[1] This accumulation of glucosylsphingosine contributes to neuronal dysfunction and destruction in patients with neuronopathic Gaucher disease[2] and it has been found to be a potent inhibitor of glucocerebrosidase. At least some instances of Gaucher disease also have a deficiency in the activity of glucopsychosine *beta*-glucosidase, the enzyme responsible for cleaving off the glucose of glucopsychosine and glucocerebroside. Like glucocerebroside and galactocerebroside, glucosylsphingosine can increase Ca_2^+ mobilization from intracellular stores although it uses a different mechanism.[3] Conduritol B epoxide (CBE), an inhibitor of *beta*-glucosidase, and l-phenyl-2-decanoylamino-3-morpholino-l-propanol (PDMP), an inhibitor of glucosylceramide synthase, can be used to create a model of Gaucher disease and consequently an accumulation of glucosylsphingosine.[4]

Selected References:

1. E. Orvisky et al. "Glucosylsphingosine accumulation in tissues from patients with Gaucher disease: correlation with phenotype and genotype" *Molecular genetics and metabolism*, Vol. 76(4) pp. 262-270, *2002*
2. R. Brady et al. "Toxicity of glucosylsphingosine (glucopsychosine) to cultured neuronal cells: a model system for assessing neuronal damage in Gaucher disease type 2 and 3" *Neurobiology of Disease*, Vol. 14(3) pp. 595-601, *2003*
3. E. Loyd-Evans et al. "Glucosylceramide and Glucosylsphingosine Modulate Calcium Mobilization from Brain Microsomes via Different Mechanisms" *The Journal of Biological Chemistry*, Vol. 278 pp. 23594-23599, *2003*
4. D. Sillance et al. "Glucosylceramide modulates membrane traffic along the endocytic pathway" *Journal of Lipid Research*, Vol. 43 pp. 1837-1845, *2002*

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