Glucocerebroside Standards for Gaucher Disease Research

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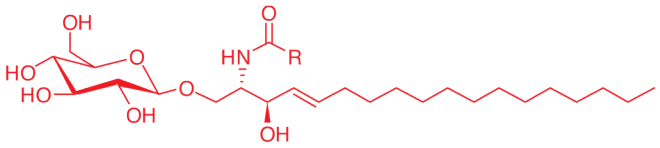

Glucocerebrosides, Gaucher's spleen
Catalog No. 1057

Glucocerebroside (glucosylceramide) is a major constituent of skin lipids where it has an important role in lamellar body formation and in maintaining the water permeability barrier. Glucocerebroside is important due to its function as the biosynthetic precursor of lactosylceramide, the backbone of many neutral oligoglycolipids and gangliosides.[1] It is found in plants, fungi, and animals and is one of the most abundant glycosphingolipids in plants. Glucocerebrosides tend to be concentrated in the outer leaflet of the plasma membrane in lipid rafts. It has been reported that glucocerebrosides are essential for the activity of tyrosinase (a key enzyme in melanin biosynthesis), for eliciting defense responses in plants, and for helping the plasma membrane in plants to withstand stresses brought about by cold and drought. Glucocerebroside has been shown to be able to modulate membrane traffic along the endocytic pathway.[2]

Gaucher disease is an inherited lysosomal storage disorder in which glucocerebroside and glucosylsphingosine accumulate in the spleen, liver, lungs, bone marrow, brain, and lymph node parenchyma due to a deficiency of the enzyme glucocerebrosidase.[3,4] Characteristic Gaucher cells are rich in accumulated glucocerebroside and glucosylsphingosine. The build up of Gaucher cells causes an enlargement of the liver and spleen. Gaucher disease affects the skeletal, hematologic, and nervous systems. Enzyme replacement therapy has proven effective for the treatment of Gaucher disease and has been shown to ameliorate or resolve the visceral aspects of type 1 Gaucher disease. The accumulation of glucocerebroside has also been associated with chemotherapy resistance.

Catalog No.	Product Name	Size	Purity
1522/1522-100	Glucocerebrosides	5 mg/100 mg	98+%
1521/1521-50	Glucocerebrosides, buttermilk	5 mg/50 mg	98+%
1057/1057-25	Glucocerebrosides, Gaucher's spleen	5 mg/25 mg	98+%
1306	Glucosylsphingosine, buttermilk	5 mg	98+%
1310	Glucosylsphingosine, plant	5 mg	98+%
2086	Glucosylsphingosine, synthetic	5 mg	98+%
2089	N-Glycinated glucosylsphingosine	1 mg	98+%
2209	$^{13}C_6$-Glucosylsphingosine	1 mg	98+%
1531	N-Docosanoyl-glucopsychosine	1 mg	98+%
1539	N-Hexanoyl-glucosylceramide	5 mg	98+%
2085	N-Hexanoyl-biotin-glucosylceramide	5 mg	98+%
1622/1622-001	N-Hexanoyl-NBD-glucosylceramide	100 µg/1 mg	98+%

References:
1. D. Sillence *et al.* (2000) Assay for the transbilayer distribution of glycolipids: selective oxidation of glucosylceramide to glucuronylceramide by TEMPO nitroxyl radicals. *Journal of Lipid Research*, Vol. 41(8) pp. 1252-1260
2. D. Sillence *et al.* (2002) Glucosylceramide modulates membrane traffic along the endocytic pathway. *Journal of Lipid Research*, Vol. 43(11) pp. 1837-1845
3. C. Walden *et al.* (2007) Accumulation of Glucosylceramide in Murine Testis, Caused by Inhibition of β-Glucosidase 2: IMPLICATIONS FOR SPERMATOGENESIS. *The Journal of Biological Chemistry*, Vol. 282(45) pp. 32655-32664
4. R. Brady (1997) Gaucher's disease: past, present and future. *Baillieres Clinical Haematology*, Vol. 10(4) pp. 621-634