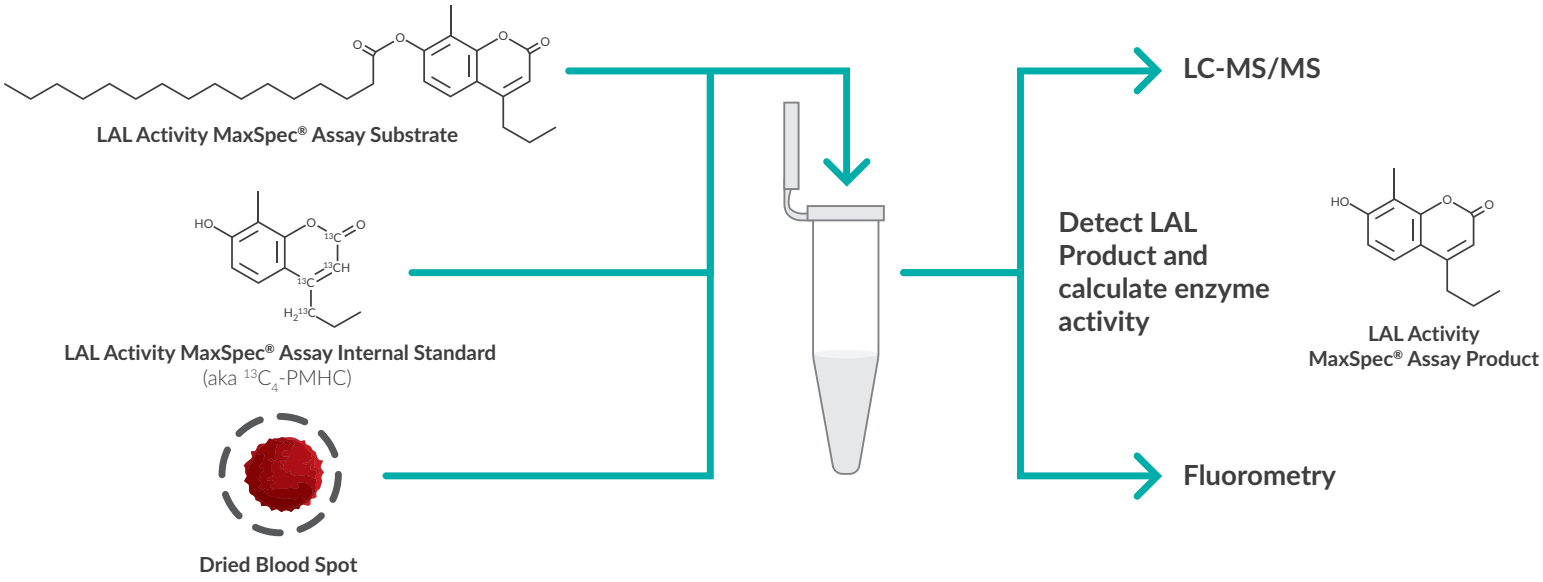
Lysosomal Storage Disorders

Lysosomal storage diseases are a group of metabolic disorders caused by the lack of key enzymes important for lysosomes to perform their normal function. While clinical trials are underway, there are few approved treatments for lysosomal storage diseases. Current research is focused on finding reliable biomarkers that can be used in these screening programs. Cayman scientists have developed LC-MS/MS assay workflows for quantitative measurement of the activity of certain key enzymes. We also offer a wide range of glycosphingolipid standards associated with the ten main sphingolipidoses that affect the glycosphingolipid pathway.

Determination of Lysosomal Acid Lipase Deficiency

Lysosomal Acid Lipase Activity MaxSpec® Assay Kit – Item No. 24854

- Easy-to-use reagent kit for the quantification of lysosomal acid lipase activity in dried blood spots
- Designed for use in LC-MS-based or fluorometry applications
- Includes necessary substrate, product, and internal standard, each provided at known concentrations



Lysosomal Acid Lipase Activity MaxSpec® Assay Kit Workflow

Related Products

Item No.	Product Name	Description
16089	4-Methylumbelliferyl Palmitate	Fluorogenic substrate for LAL
23891	Lalistat 1	An inhibitor of LAL (IC_{50} = 68 nM)
25347	Lalistat 2	An inhibitor of LAL (IC_{50} = 152 nM)
601810	Lysosomal Staining Kit (Red Fluorescence)	For imaging of lysosomes in live cells
25152	LysoBrite™ Blue	A lysosomal dye
25154	LysoBrite™ Green	A lysosomal dye
25157	LysoBrite™ Red	A lysosomal dye

Learn more at **www.caymanchem.com/lysosomalstorage**

Sphingolipid Biomarkers

Sphingolipidoses (sphingolipid lysosomal storage disorders) result in an accumulation of various sphingolipids in the lysosome. Ten main sphingolipidoses affect the glycosphingolipid pathway: Farber, Krabbe, Gaucher, Metachromatic Leukodystrophy, Fabry, Sandhoff, Niemann-Pick, Sialidosis, Tay-Sachs, and G_{M1} gangliosidosis. All of these disorders are characterized by an accumulation of sphingolipids in the lysosome due to enzyme deficiency or ineffective transport of lipids from the lysosme. The schematics below detail the enzymes associated with these disorders and the sphingolipid products they produce.

Sphingosine and Ceramide Accumulation

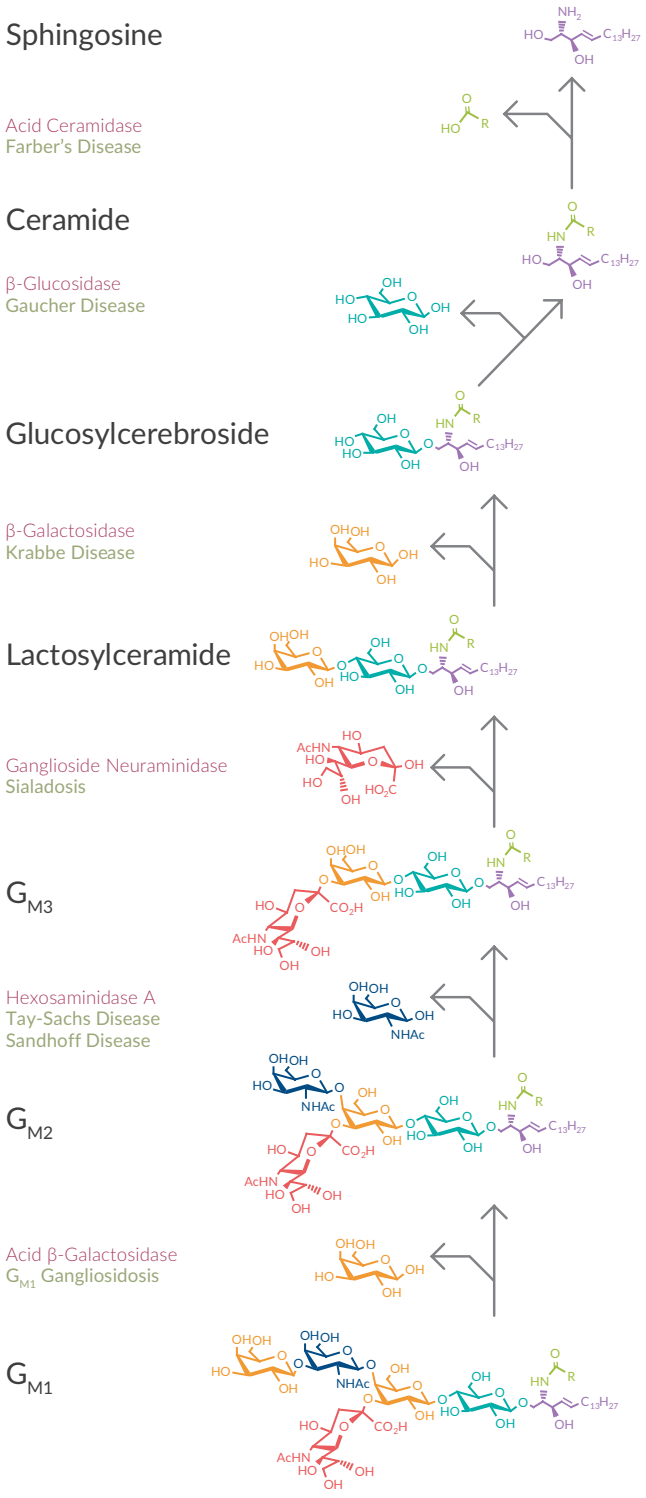
Item No.	Product Name
9000415	C6 Biotin Ceramide (d18:1/6:0)
22532	C17 Ceramide (d18:1/17:0)
24396	C18 Ceramide-d_3 (d18:1/18:0-d_3)
24428	C18 dihydro Ceramide-d_3 (d18:0/18:0-d_3)
24464	C18 Ceramide-1-Phosphate-d_3 (d18:1/18:0-d_3)
10007907	Sphingosine (d18:1)
24371	Sphingosine-d_9 (d18:1)

Glucosylceramide Accumulation

Item No.	Product Name
23206	Glucocerebrosides (buttermilk)
23207	Glucocerebrosides (Gaucher's spleen)
25850	Glucocerebrosides (soy)
24621	C16 Glucosylceramide-d_3 (d18:1/16:0-d_3)
23208	C6 Biotin Glucosylceramide (d18:1/6:0)
23209	C6 NBD Glucosylceramide (d18:1/6:0)
23213	1-β-D-Glucosylsphingadienine (d18:2 (4E,8E))
23212	$^{13}C_6$ Glucosylsphingosine (d18:1)
23211	1-β-D-Glucosylsphingosine (d18:1)
24473	N-Glycine Glucosylsphingosine (d18:1)

Galactosylceramide Accumulation

Item No.	Product Name
24471	C6 Galactosylceramide-biotin
22830	C6 NBD Galactosylceramide (d18:1/6:0)
24466	C15 Galactosylceramide (d18:1/15:0)
22851	C12 NBD Galactosylceramide (d18:1/6:0)
24467	C18 Galactosylceramide-d_{35} (d18:1/18:0-d_{35})
24322	Galactosylcerebrosides (bovine)
20338	Galactosylsphingosine (d18:1)
24620	N-Glycine Galactosylsphingosine (d18:1)



Lipid Class · Enzyme Deficiency · Associated Disorder

Gangliosides Accumulation

Item No.	Product Name
24852	Ganglioside G_{D3}-d_3
27202	C6 Biotin Ganglioside G_{D3} (d18:1/6:0)
24840	C6 Biotin Ganglioside G_{M1} (d18:1/6:0)
24839	C18 Ganglioside G_{M1}-d_3 (d18:1/18:0-d_3) (ammonium salt)
24837	Lyso-Monosialoganglioside G_{M1} (ammonium salt)
24849	C18 Ganglioside G_{M2}-d_3 (d18:1/18:0-d_3) (ammonium salt)
24850	C18 Ganglioside G_{M3}-d_3 (d18:1/18:0-d_3) (ammonium salt)

Lactosylceramides Accumulation

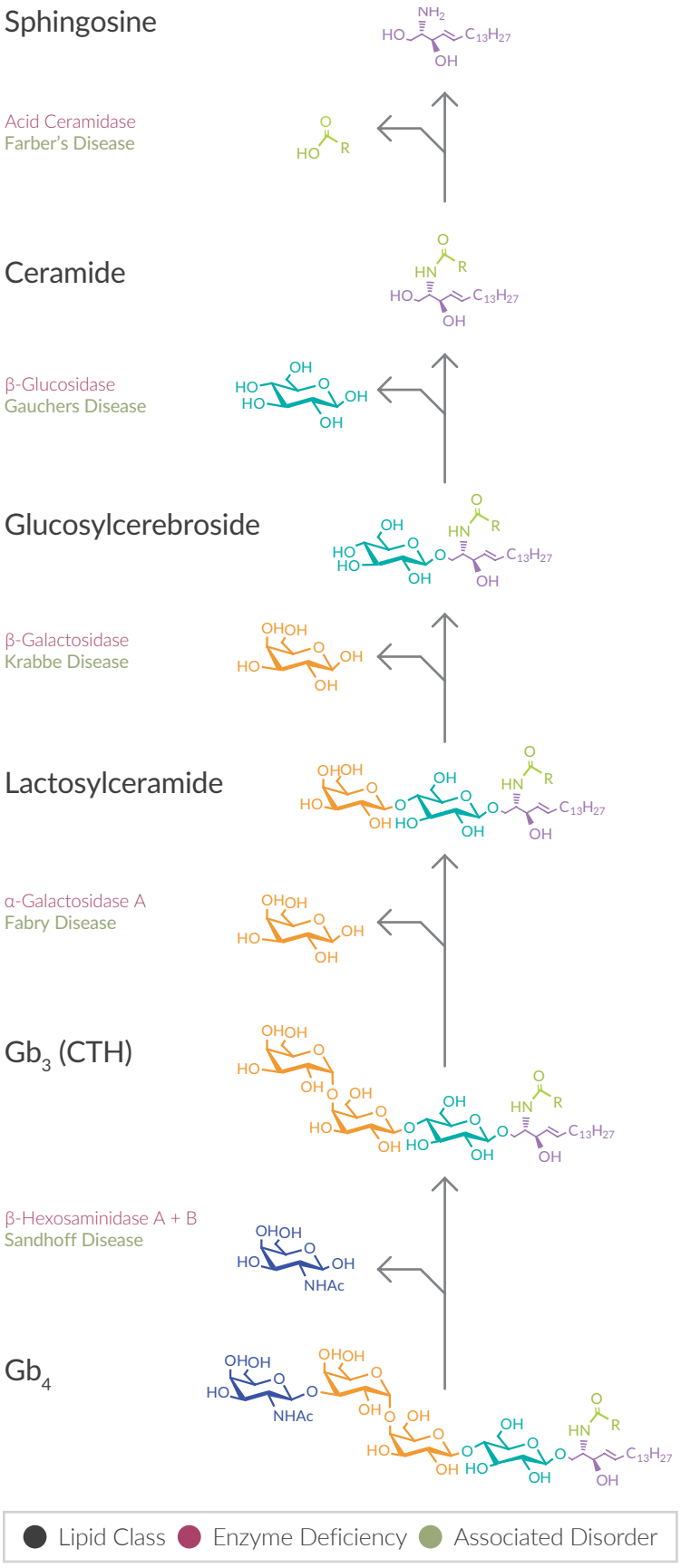
Item No.	Product Name
16983	Lactosylceramides (bovine brain)
27197	Lactosylceramides (bovine buttermilk)
24859	C6 Biotin Lactosylceramide (d18:1/6:0)
22828	C6 NBD Lactosylceramide (d18:1/6:0)
22829	C12 NBD Lactosylceramide (d18:1/12:0)
24625	C16 Lactosylceramide-d_3 (d18:1/16:0-d_3)
24868	Lactosylsphingosine (d18:1)
24867	Lactosylsphingosine (d18:1) (synthetic)
24869	N-Glycine Lactosylsphingosine (d18:1)

Ceramide Trihexosides (Globotriaosylceramides) Accumulation

Item No.	Product Name
24870	Globotriaosylceramides (porcine)
24873	Lyso-Globotriaosylceramide (d18:1)
24628	C12 NBD Globotriaosylceramide (C18:1/12:0)
24876	C17 Globotriaosylceramide (d18:1/17:0)
24626	C18 Globotriaosylceramide-d_3 (d18:1/18:0-d_3)
24874	N-Glycine Globotriaosylsphingosine (d18:1)

Globosides (Globotetrahexosylceramides) Accumulation

Item No.	Product Name
24881	Globotetraosylceramides (porcine RBC)

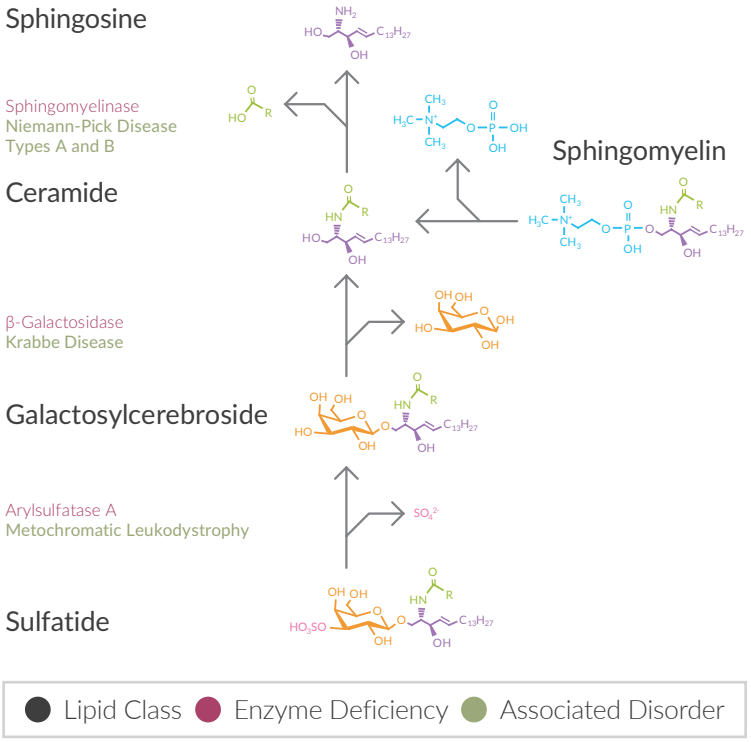


Sphingosylphosphorylcholine Accumulation

Item No.	Product Name
24328	C6 NBD Sphingomyelin (d18:1/6:0)
24329	C12 NBD Sphingomyelin (d18:1/12:0)
10007946	C16 Sphingomyelin (d18:1/16:0)
24452	^{13}C C16 Sphingomyelin (d18:1/16:0)
24354	C17 D-*erythro*/L-*threo* Sphingomyelin (d18:1/17:0)

Sulfatides Accumulation

Item No.	Product Name
24323	Sulfatides (bovine) (sodium salt)
24624	C18 3'-sulfo Galactosylceramide-d_3 (d18:1/18:0-d_3)
24627	C12 NBD 3'-sulfo Galactosylceramide (d18:1/12:0)
25316	3'-sulfo Galactosylsphingosine (ammonium salt)
27200	N-Glycine 3'-sulfo Galactosylsphingosine
24858	C6 Biotin 3'-sulfo Galactosylceramide (d18:1/6:0)



View a complete list of our glycosphingolipids at **www.caymanchem.com**