Sphingolipidosis|: are group of inherited disorders of sphingolipids and glycolipids due to genetic deficiency of one or more of enzymes involved in degradetion of these types of lipids with a range of clinical features.

Disease	Defective Enzyme[a]	Accumulated Intermediate
GM_1 gangliosidosis	1 β-Galactosidase	GM_1 ganglioside
Tay–Sachs disease	2 β-*N*-Acetylhexosaminidase A	GM_2 (Tay–Sachs) ganglioside
Fabry's disease	3 α-Galactosidase A	Trihexosylceramide
Gaucher's disease	4 β-Glucosidase	Glucosylceramide
Niemann–Pick disease	5 Sphingomyelinase	Sphingomyelin
Farber's lipogranulomatosis	6 Ceramidase	Ceramide
Globoid cell leukodystrophy (Krabbe's disease)	7 β-Galactosidase	Galactosylceramide
Metachromatic leukodystrophy	8 Arylsulfatase A	3-Sulfogalactosyl-ceramide
Sandhoff disease	9 *N*-Acetylhexosaminidases A and B	GM_1 ganglioside and globoside

[a]Numbers refer to enzymes shown in Figure 19.16.