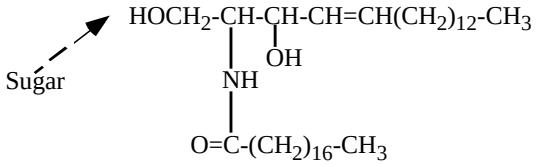
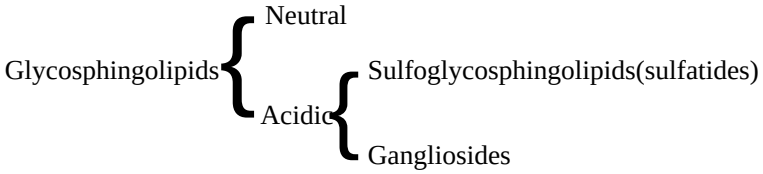
GLYCOSPHINGOLIPIDS

General Structural Features:



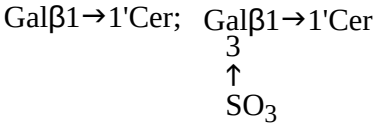
Structure of a ceramide: N-stearoylsphingosine

Classification:

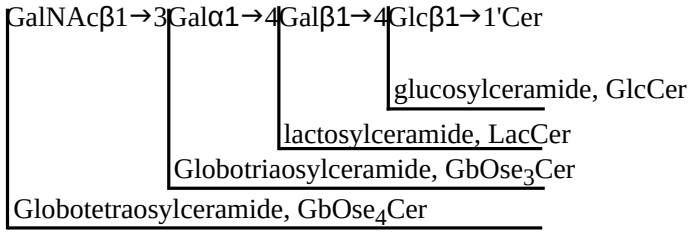


Structures of Glycosphingolipids Commonly Found in Higher Animals:

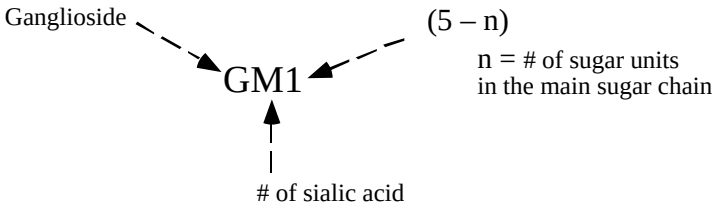
1) *Glycosphingolipids of GalCer Family*



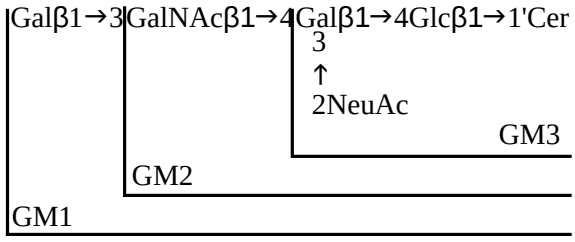
2) *Major Neutral Glycosphingolipids Found In Higher Animals*



3) *Svennerholm's Nomenclature for Gangliosides*



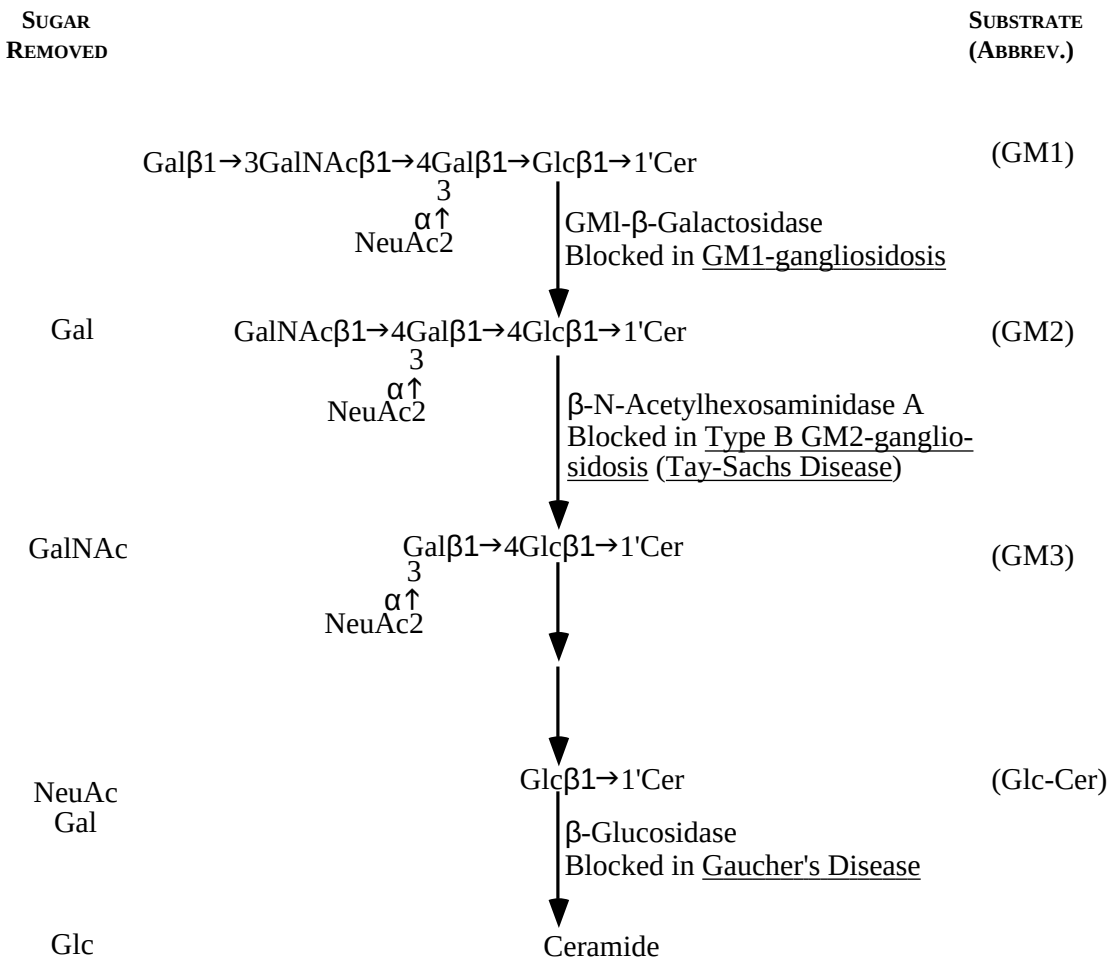
4) *Major Gangliosides Found in Higher Animals*



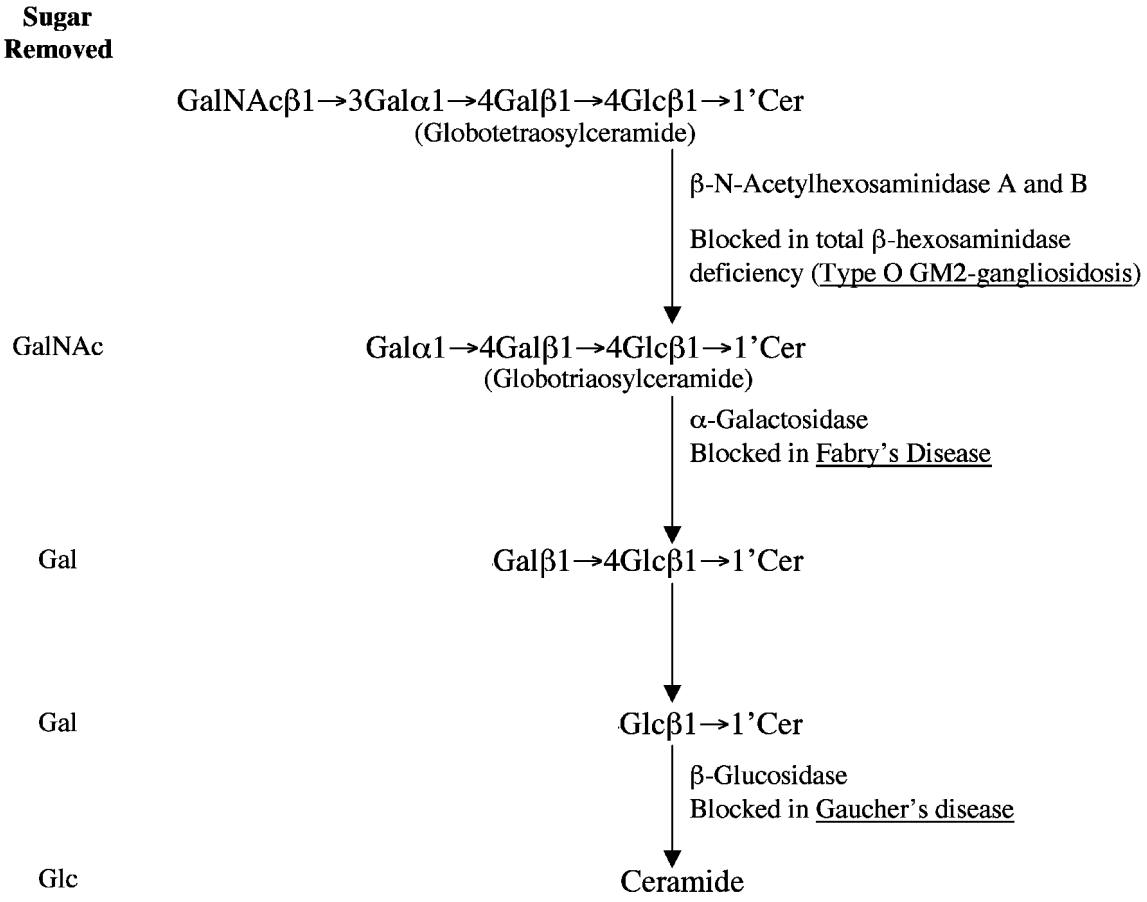
INBORN ERRORS OF GLYCOSPHINGOLIPID CATABOLISM

Glycosphingolipid Storage Diseases (also called **Sphingolipidoses**) are congenital disorders characterized by excessive accumulation of glycosphingolipids in the tissues of the patient, due to the impaired degradation of a specific glycosphingolipid. Recently it has been shown that in addition to glycosidases the catabolism of glycosphingolipids requires protein cofactors called activator proteins. The following two schemes show the relationship between the degradation of glycosphingolipids and several sphingolipidoses.

SCHEME 1. **CATABOLISM OF GM1-GANGLIOSIDE**



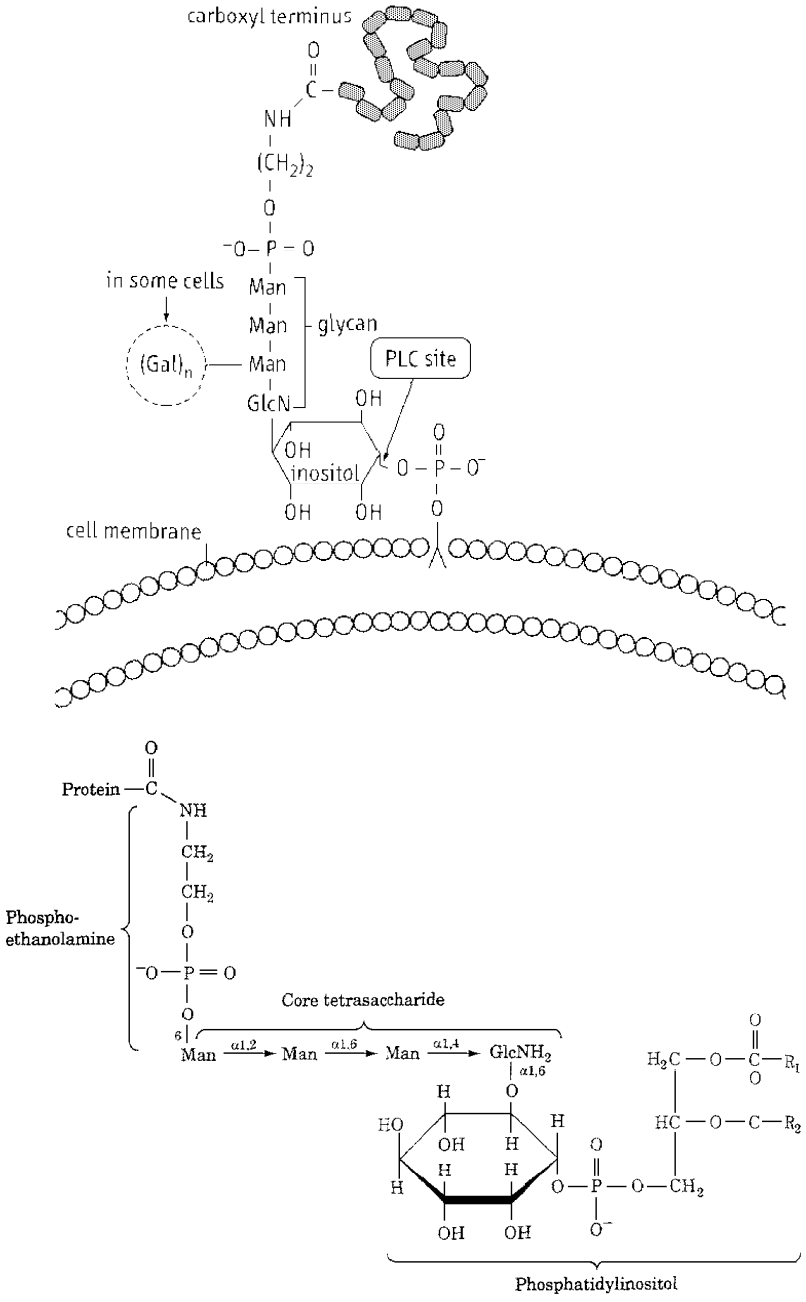
SCHEME 2. **CATABOLISM OF GLOBOTETRAOSYLCERAMIDE**



GLYCOSYLPHOSPHATIDYLINOSITAL (GPI) ANCHORS OF PROTEINS

Glycosylphosphatidylinositol-linked proteins (GPI-linked or GPI-anchored proteins) occur in eucaryotes, but are particularly abundant in some parasitic protozoa, which contain relatively few membrane proteins anchored by transmembrane polypeptide segments. Like glycoproteins and glycolipids, GPI-anchored proteins are located only on the exterior surface of the plasma membrane.

GPI anchor and protein attachment



The core structure of the GPI anchors of proteins.