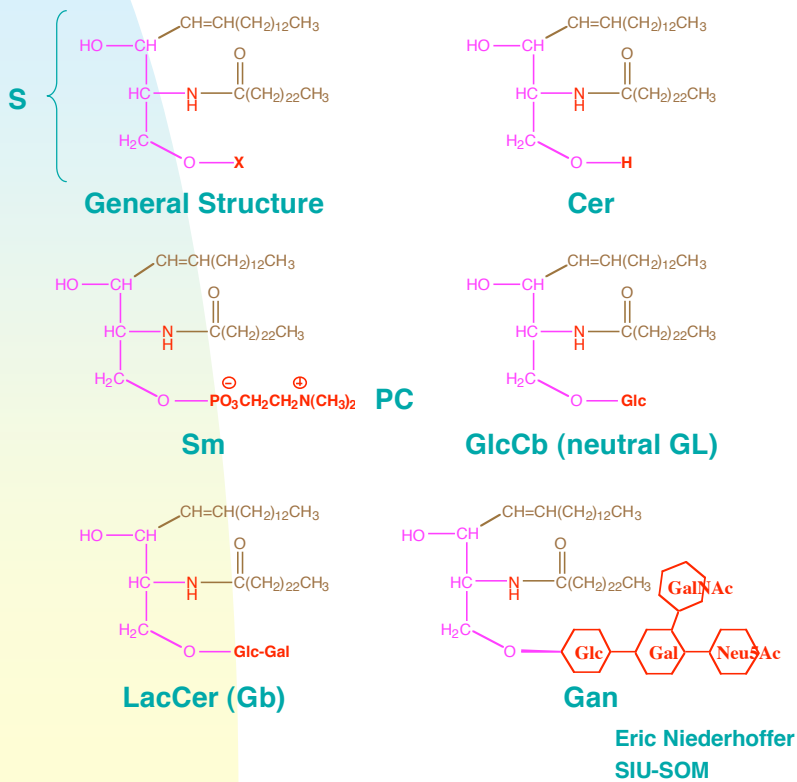


Sphingolipid Disorders

Sphingolipids (phospho- or glycolipids)



S: sphingosine

Cer: ceramide

Sm: sphingomyelin

PC: phosphocholine

Glu: glucose

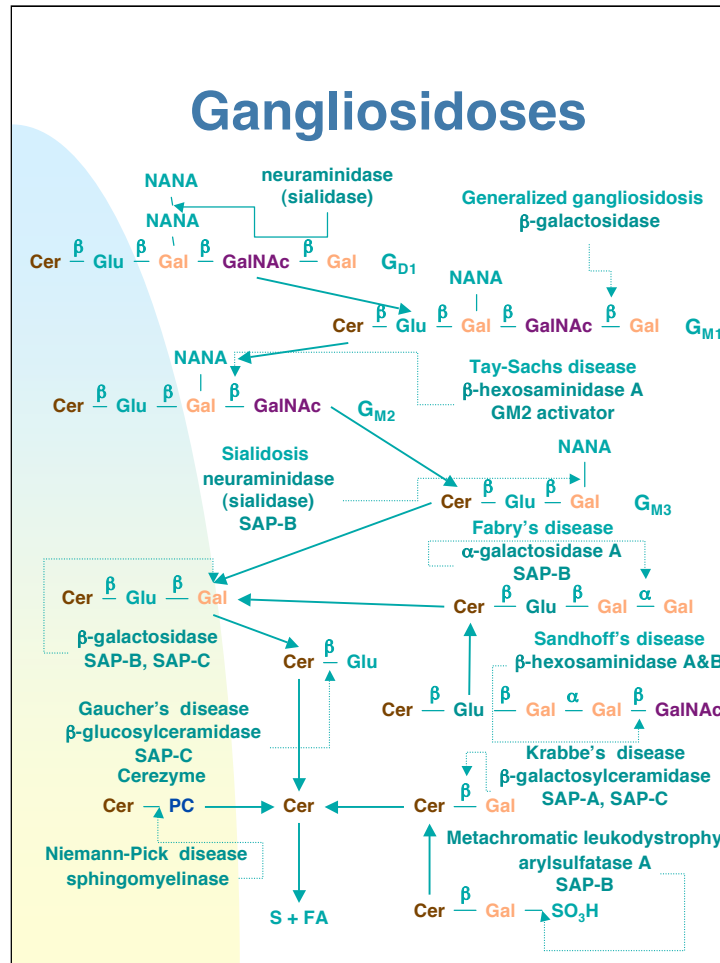
GL: glycolipid

Lac: lactose

Gb: globoside (two or more sugars plus GalNAc)

Gan: ganglioside (contains NANA)

Cb: cerebroside (single sugars)



NANA: *N*-acetylneuraminic acid (sialic acid)

Cer: ceramide **Glu:** glucose **Gal:** galactose

GalNAc: *N*-acetylgalactosamine **G:** ganglioside

D1: dineuraminic acid + Gal-GalNAc-Gal-Glc-Cer

M1: neuraminic acid + Gal-GalNAc-Gal-Glc-Cer

M2: neuraminic acid + GalNAc-Gal-Glc-Cer

M3: neuraminic acid + Gal-Glc-Cer

SAP: sphingolipid activator protein (saposin)

Cer-Glu-Gal: lactosylceramide **Cer-Glu:** glucosylceramide

S: sphingosine **FA:** fatty acid

Cer-Glu-Gal-Gal-GalNAc: globoside **Cer-Glu-Gal-Gal:** globotriaosylceramide

PC: phosphocholine **Cer-PC:** sphingomyelin

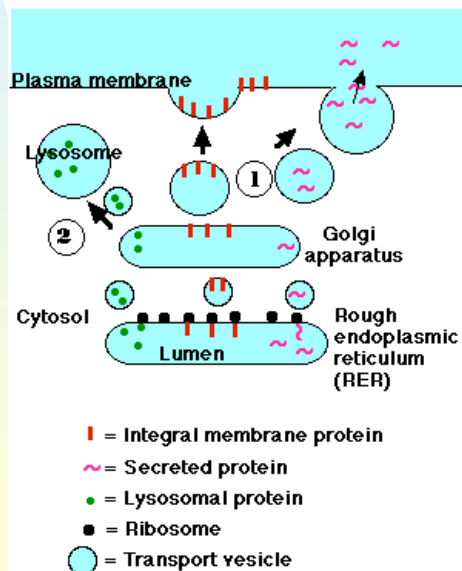
Cer-Gal-SO₃H: sulfatide **Cer-Gal:** galactosylceramide

Targeting of Lysosomal Enzymes to Lysosomes

Addition of M6P to lysosomal enzymes

Recognition by MPRs

M6P independent pathways



M6P: mannose-6-phosphate

MPR: mannose-6-phosphate receptor

Review Questions

- **How do you interpret ganglioside names (G, D, M, 1, 2, 3)?**
- **What do the different lysosomal enzyme names mean in the context of removing saccharides?**
- **Where does ganglioside degradation occur?**