



Medical Committee
The University of Jordan



SLIDE



SHEET

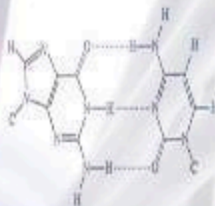


SLIDE : 19



DR.NAME: Faisal Al-Khateib

Biochemistry



Majida Al-Foqaraa'

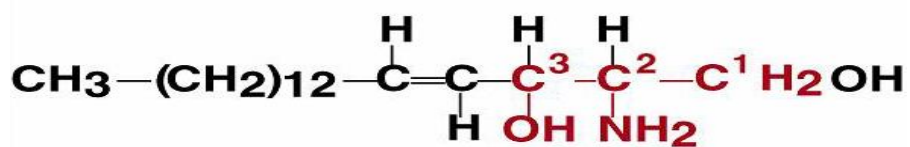
Metabolism of Sphingolipids

Sphingophospholipids

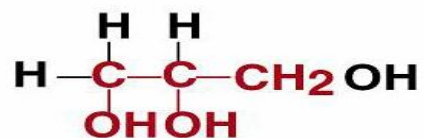
Glycosphingolipids

Faisal Khatib MD; PhD
Faculty of Medicine, University
of Jordan

Sphingosine; Amino Alcohol

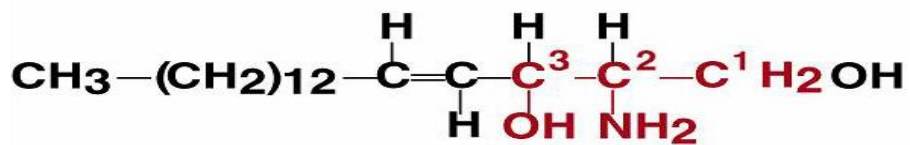


Sphingosine

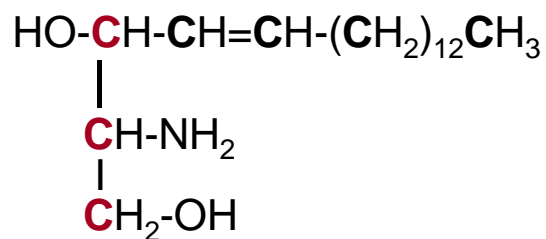


Glycerol

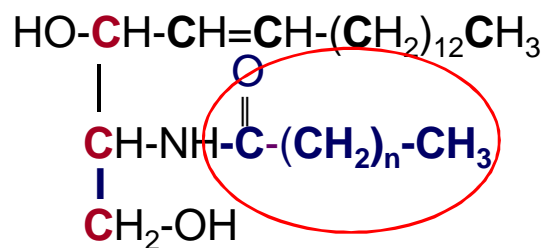
Sphingosine; Amino Alcohol



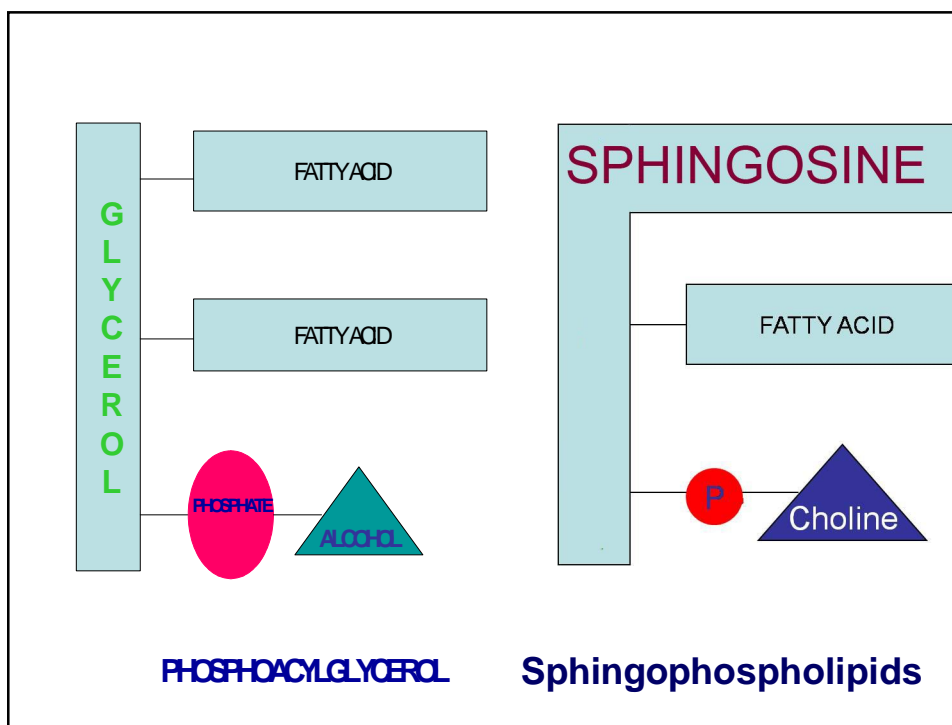
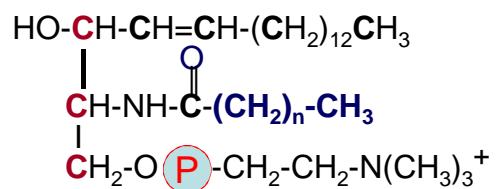
Sphingosine

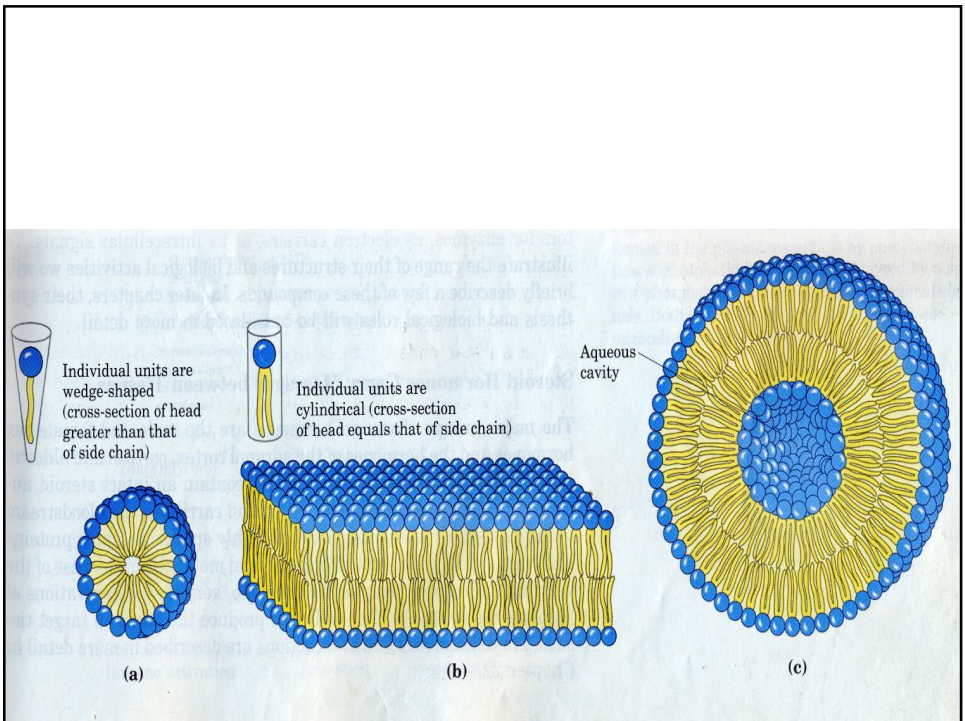
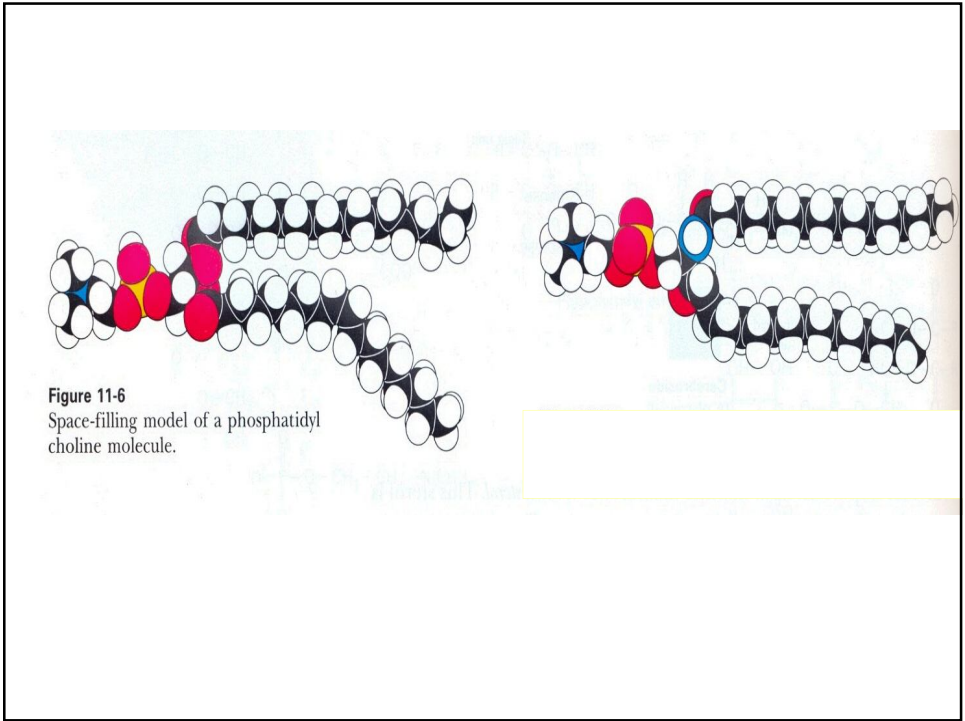


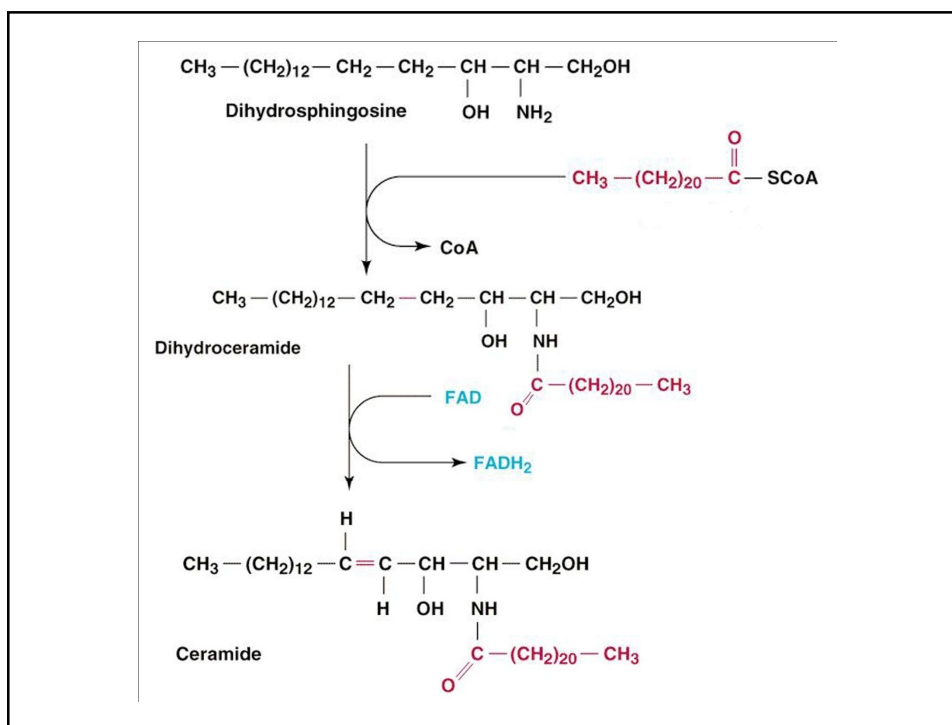
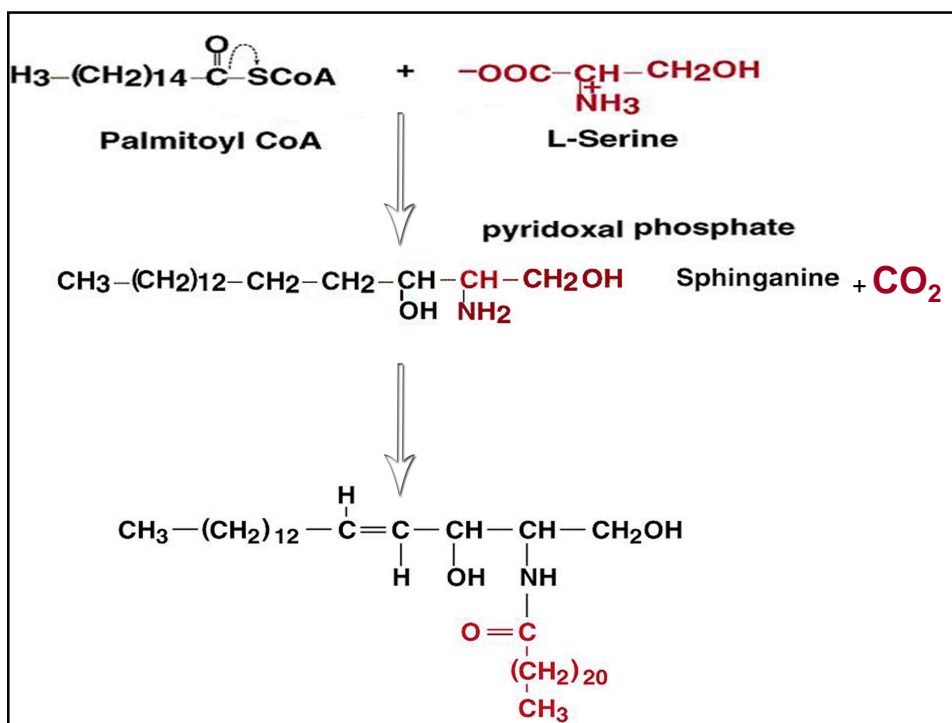
Ceramide: Fatty Acid to joined to Sphingosine

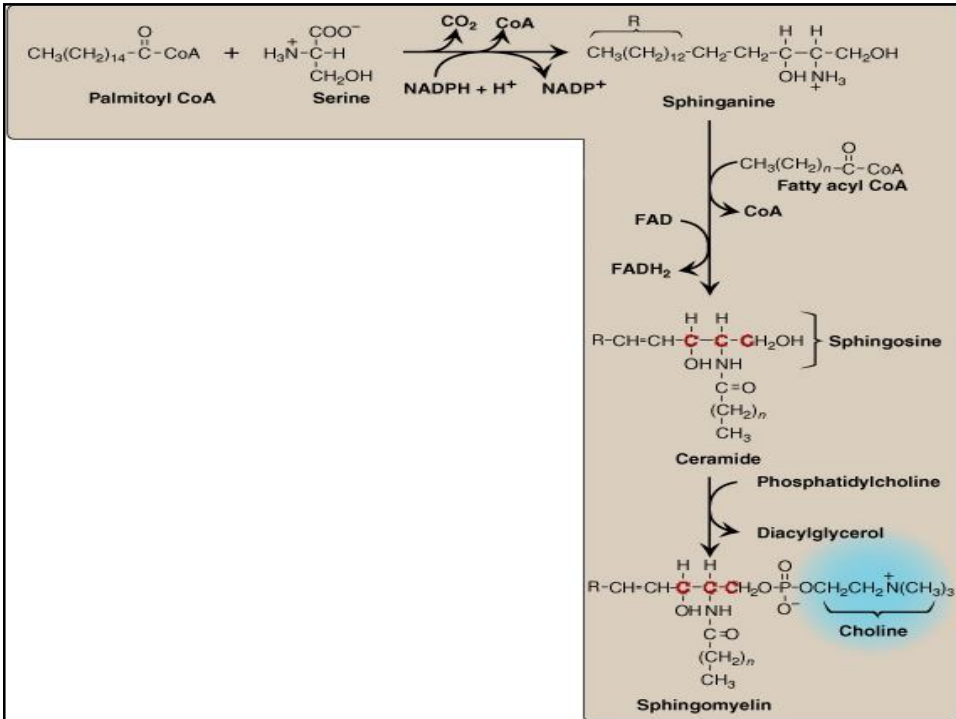
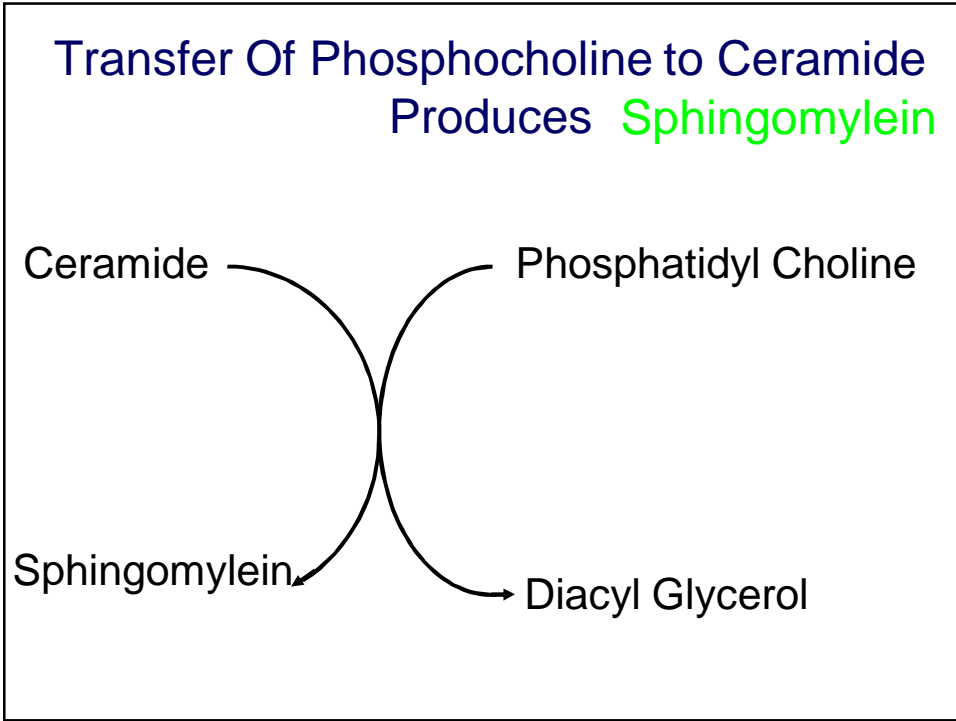


Spingomyelin is Phosphocholine Ester of Ceramide







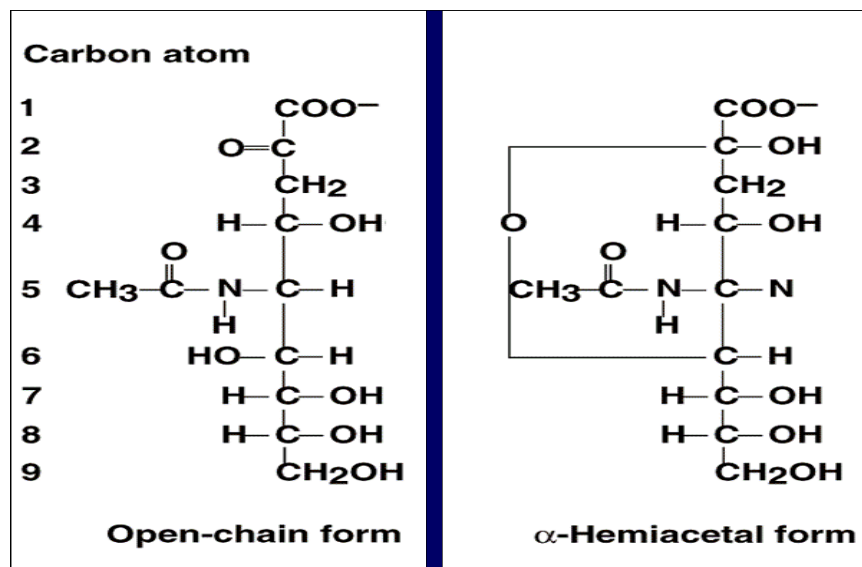


Glycolipids are Formed by Linking one or More Sugars to Ceramide

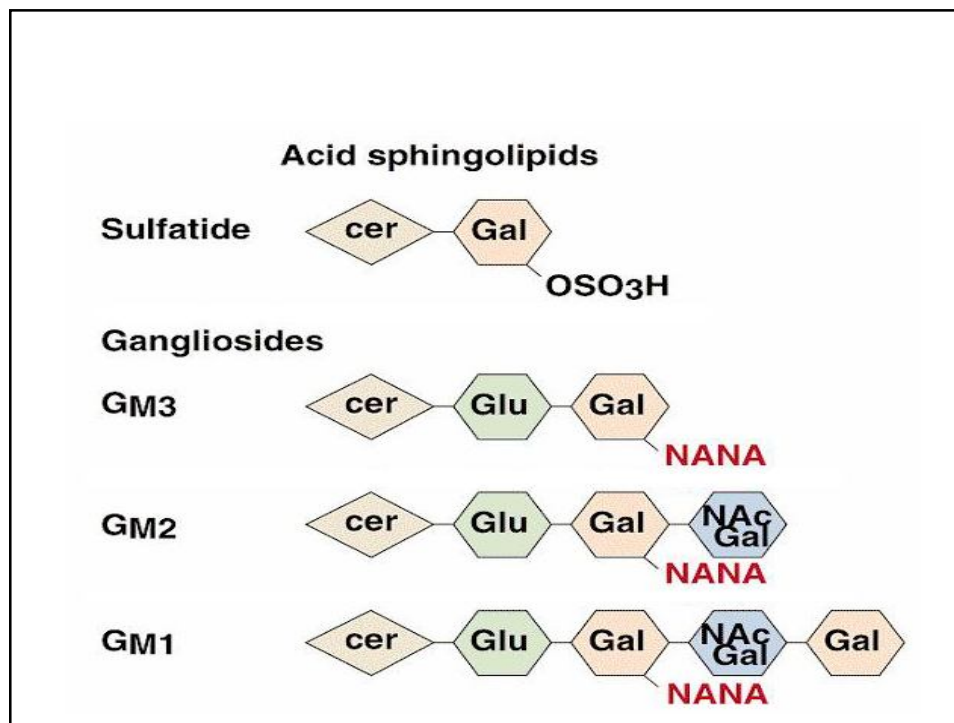
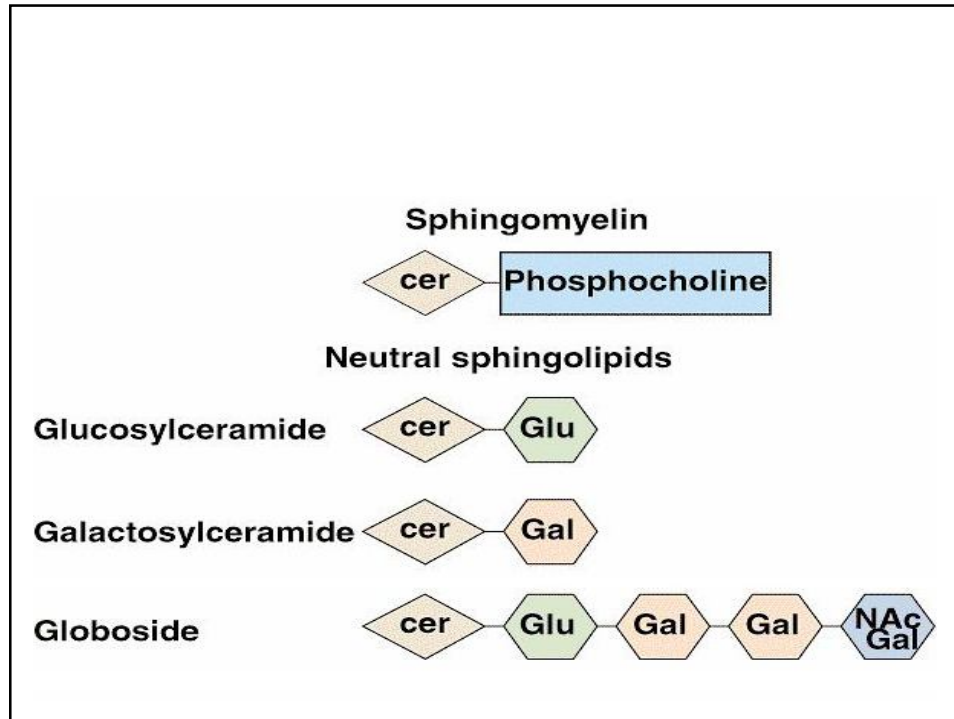
Ceramide +

- Glucose or Galactose => **Cerebroside**
- Sulfated Galactose => **Sulfoglycosphingolipids**
- Oligosaccharide => **Globoside**
- Oligosaccharide with NANA => **Gangliosides**

N-Acetylneuraminic Acid (NANA)

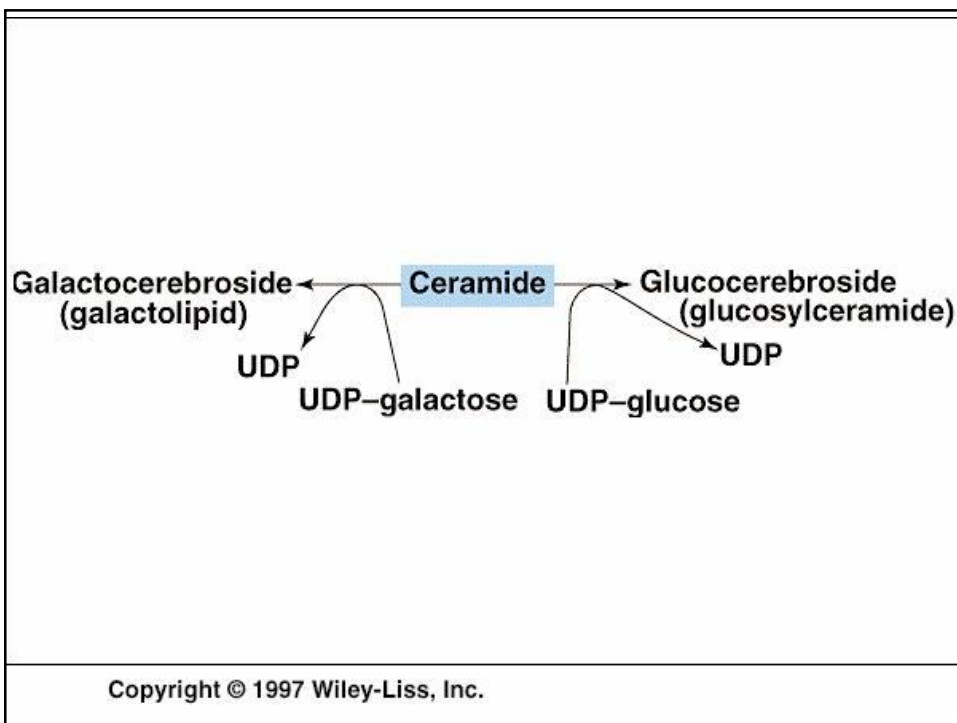


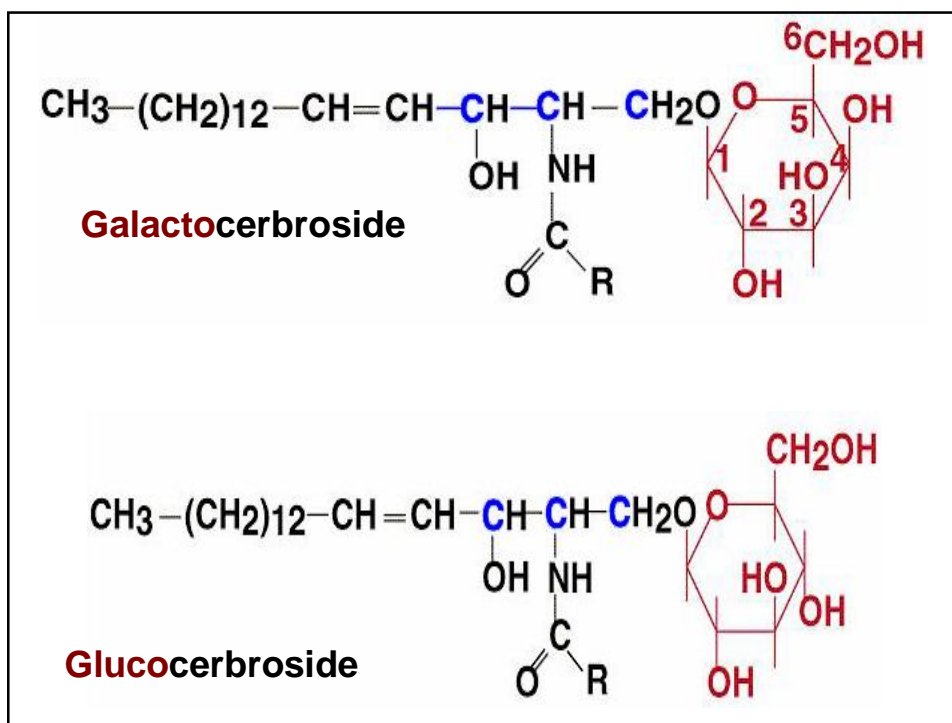
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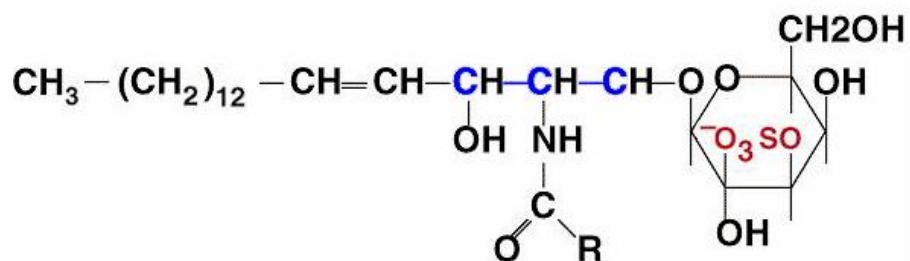
Activated Donors in Glycolipids Synthesis

- UDP-**Glucose**
- UDP-**Galactose**
- UDP-**N-Acetylgalctoseamine**
- CMP- **N-Acetylneuraminic Acid**

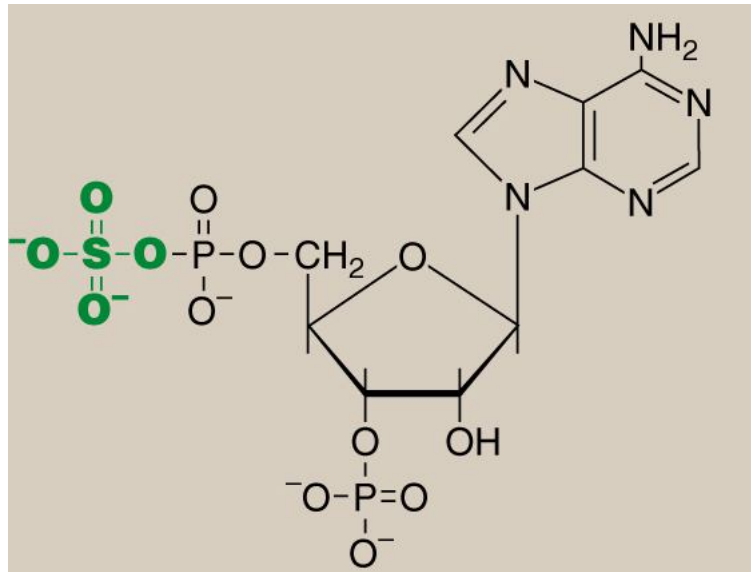




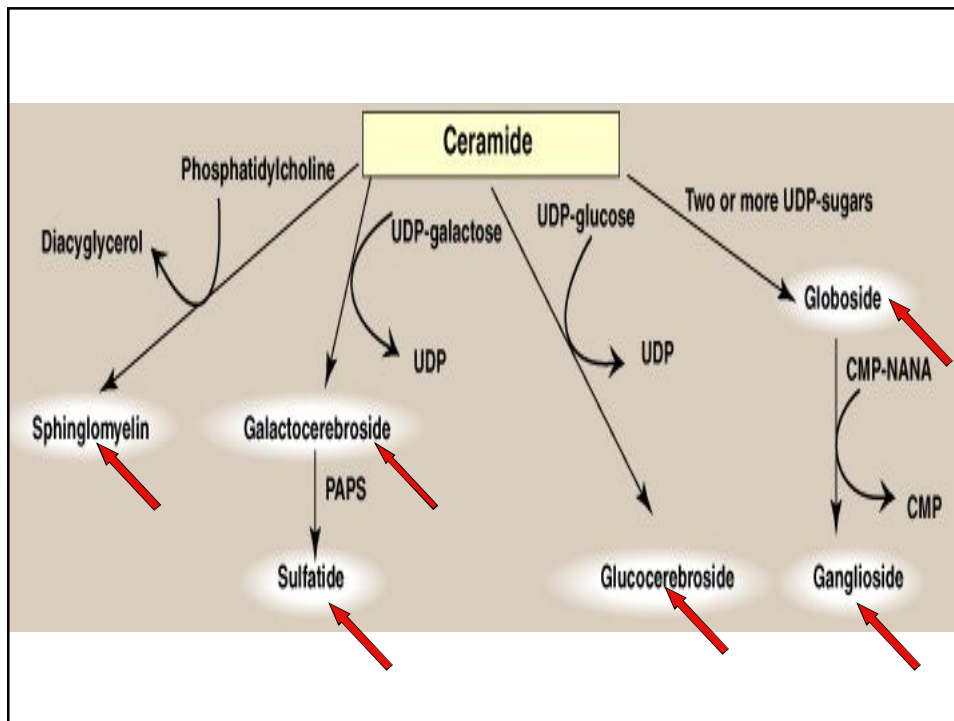
Transfer of Sulfate Group to Galactocerebroside Produces **Sulfogalactocerebroside** (Sulfatide)



Sulfate Group Donor

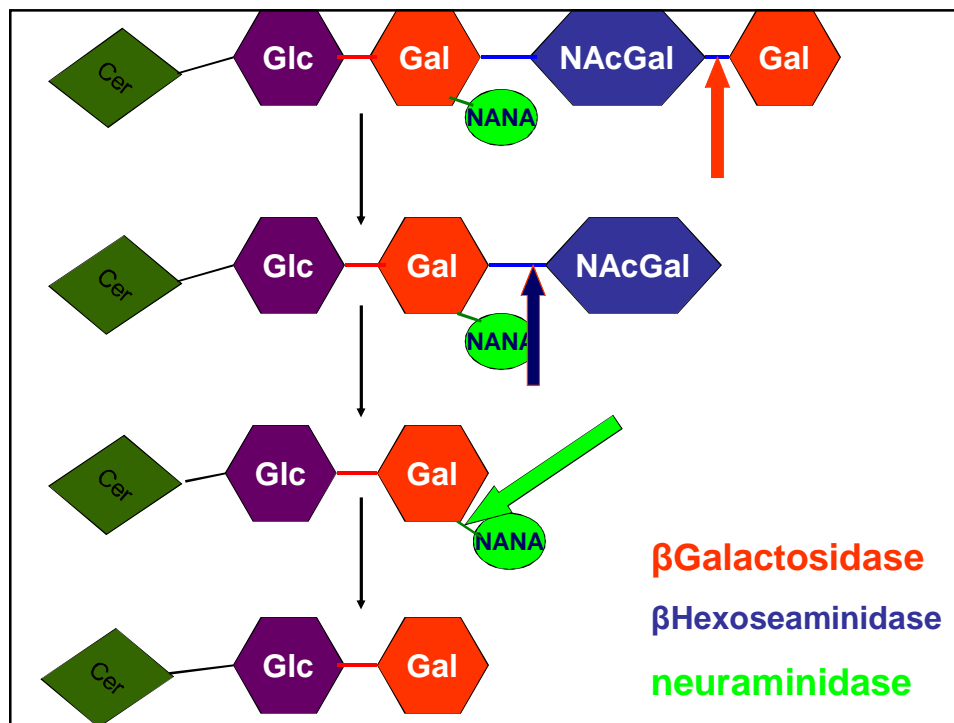


3 Phosphoadenosine 5 Phosphosulfate PAPS



Degradation of Sphingolipids

- Hydrolytic Enzymes, Specific for the Sugar
 - - α Galactosidase
 - - β Galactosidase
 - - neuraminidase
 - - Hexoaminidase
- In Lysosomes
- Enzymes are firmly Bound to Lysosomal Membrane.
- The pH Optimum 3.5-5.5
- Stepwise Sequential Process
- “Last on, First off”



Sphingolipidoses

- Lipid Storage Diseases
- **Defect in one of the Enzyme**
- Inherited as Autosomal Recessive Disease
- **Accumulation of Specific Lipid**
Substrate of the Defective Enzyme
- **Brain is Mostly Affected.**
- Extent of Enzyme deficiency is the same in Different Tissues.

Degradation of Sphingomyelin

