

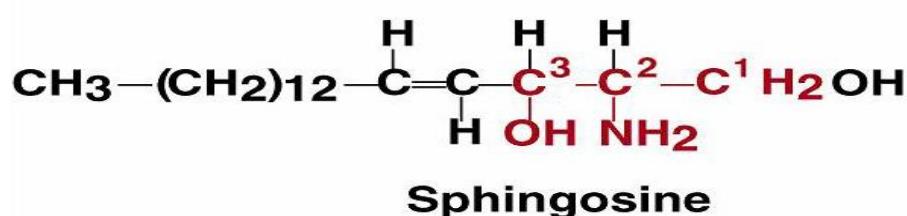
# Metabolism of Sphingolipids

## Sphingophospholipids

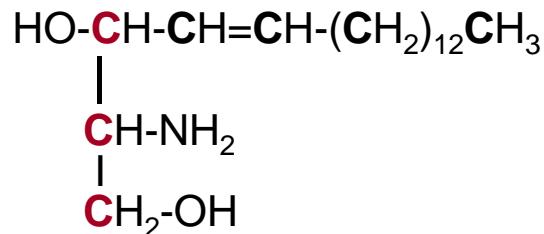
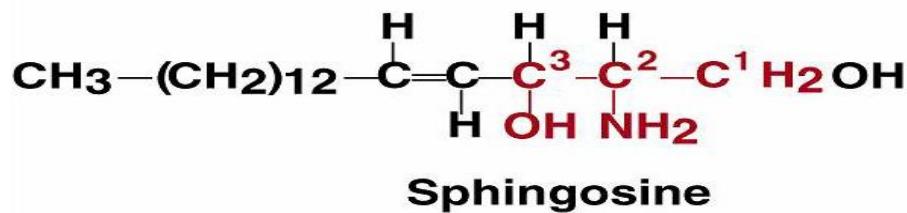
## Glycosphingolipids

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Faculty of Medicine, University  
of Jordan

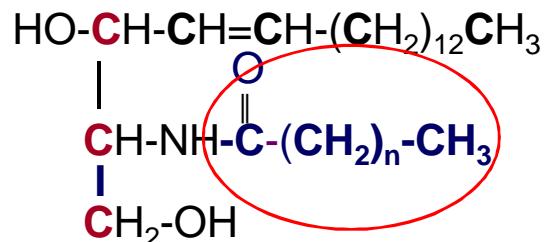
### Sphingosine; Amino Alchol



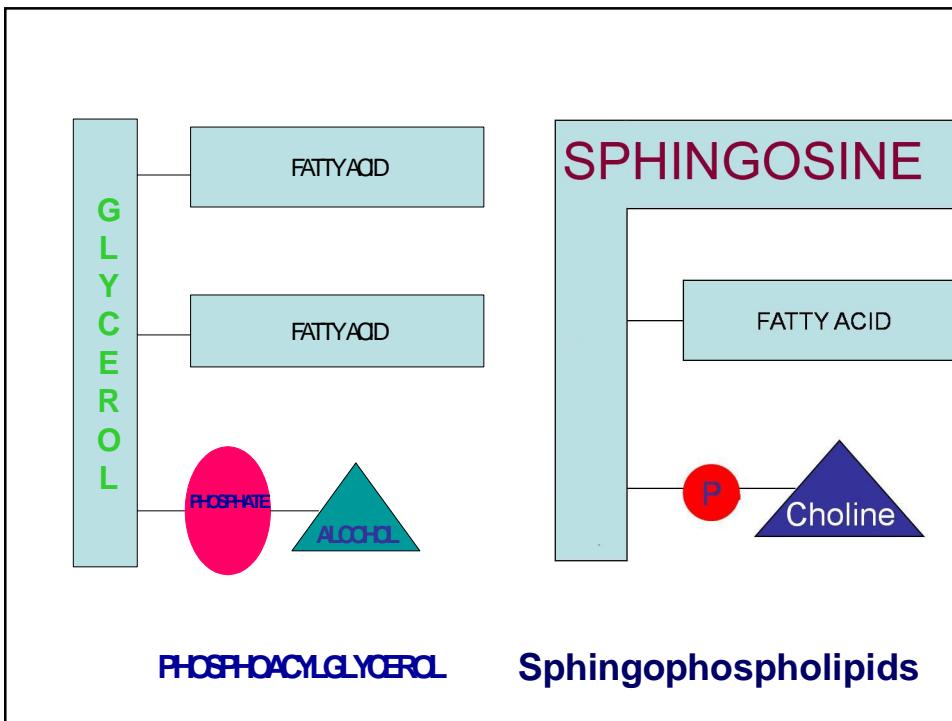
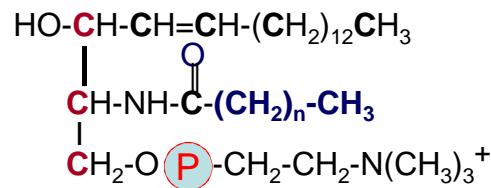
## Sphingosine; Amino Alchol

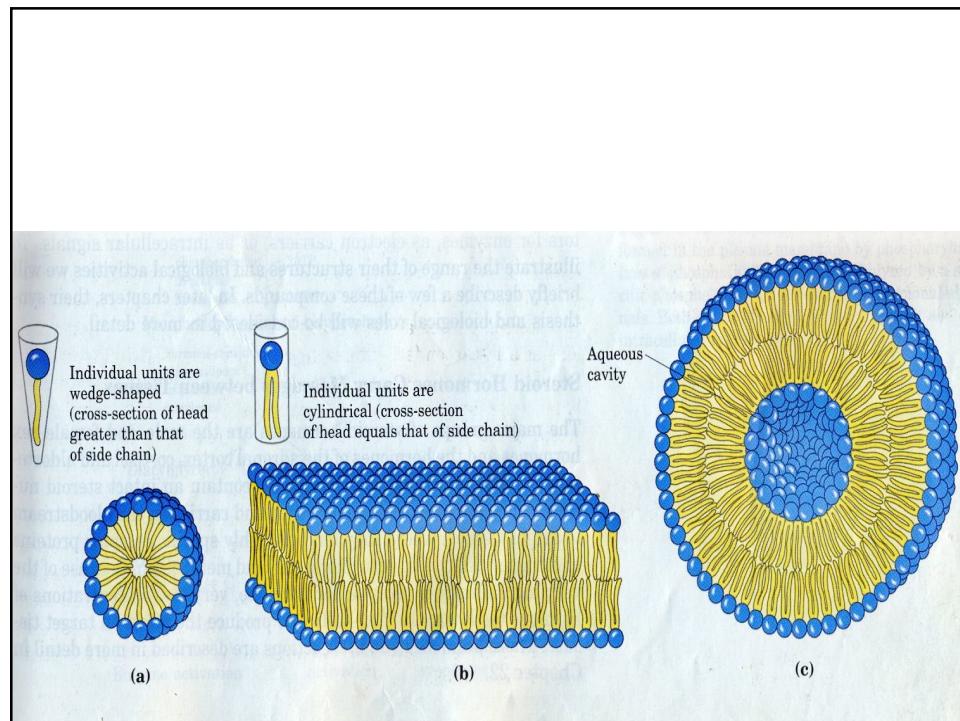
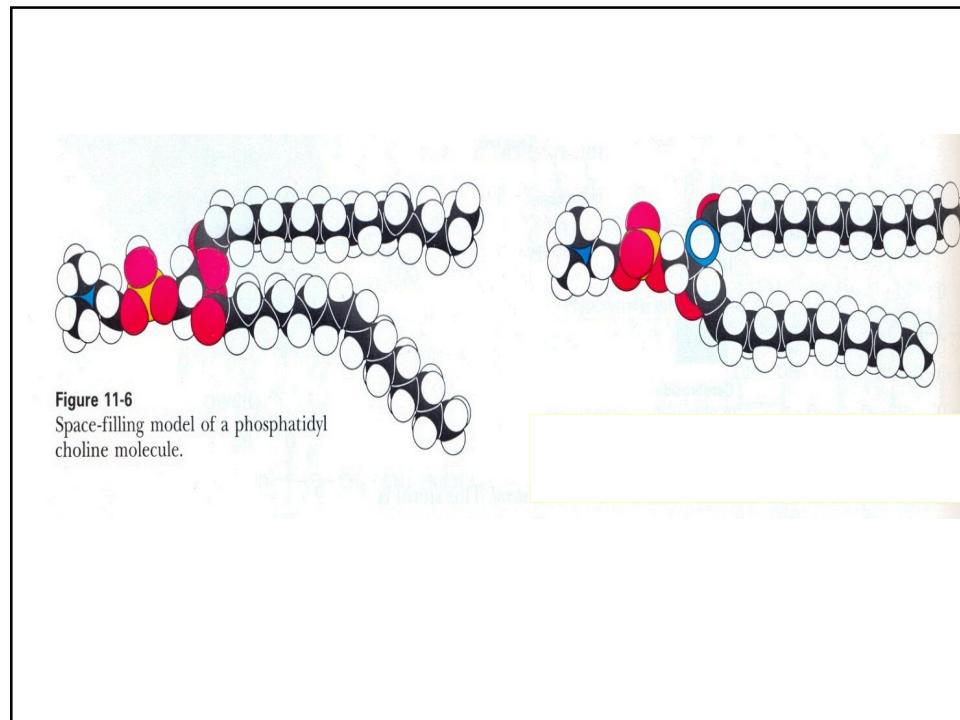


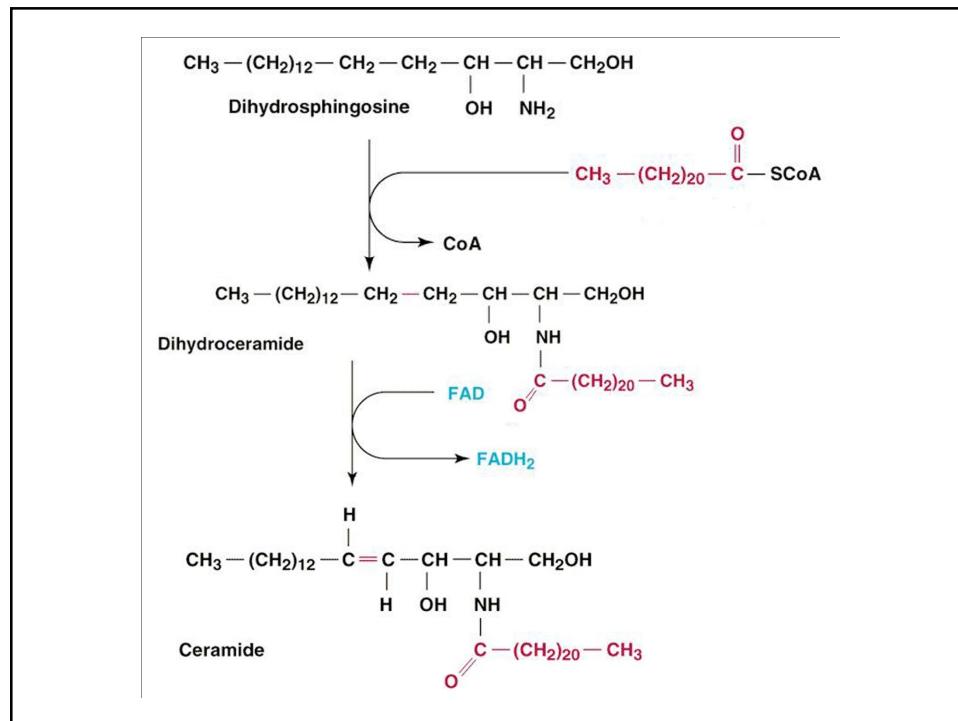
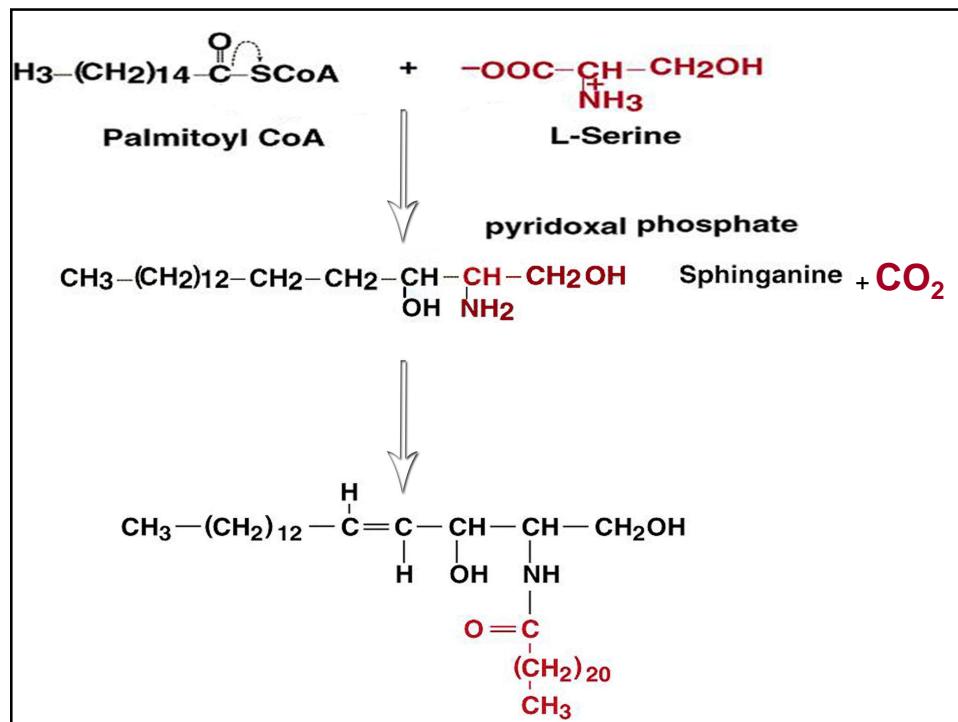
## Ceramide: Fatty Acid to joined to Sphingosine



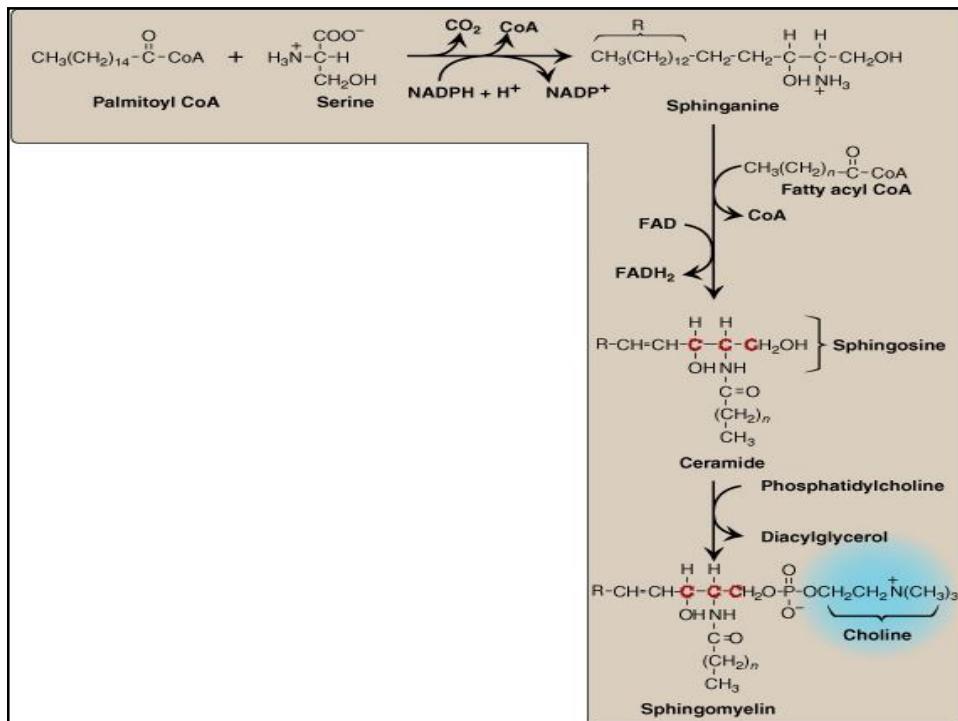
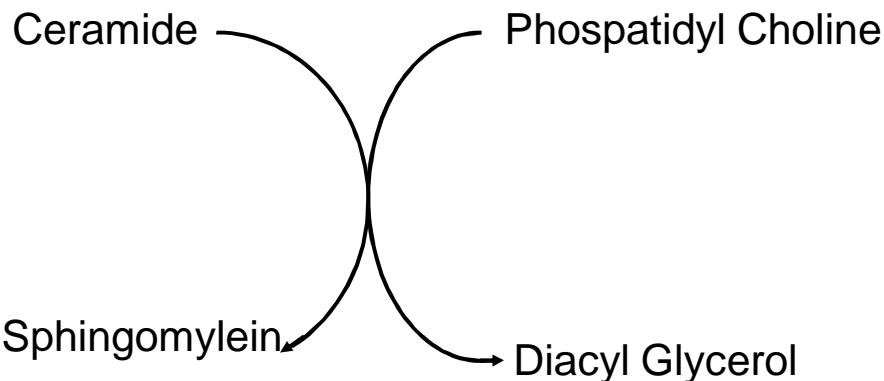
## Sphingomyelin is Phosphocholine Ester of Ceramide







## Transfer Of Phosphocholine to Ceramide Produces Sphingomylein



## Glycolipids are Formed by Linking one or More Sugars to Ceramide

### Ceramide +

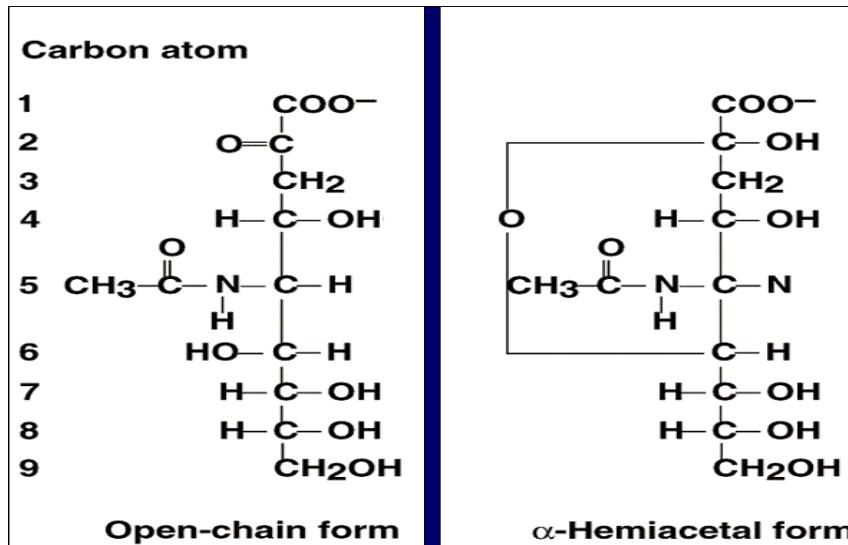
-Glucose or Galactose => Cerebroside

-Sulfated Galactose => Sulfoglycosphingolipids

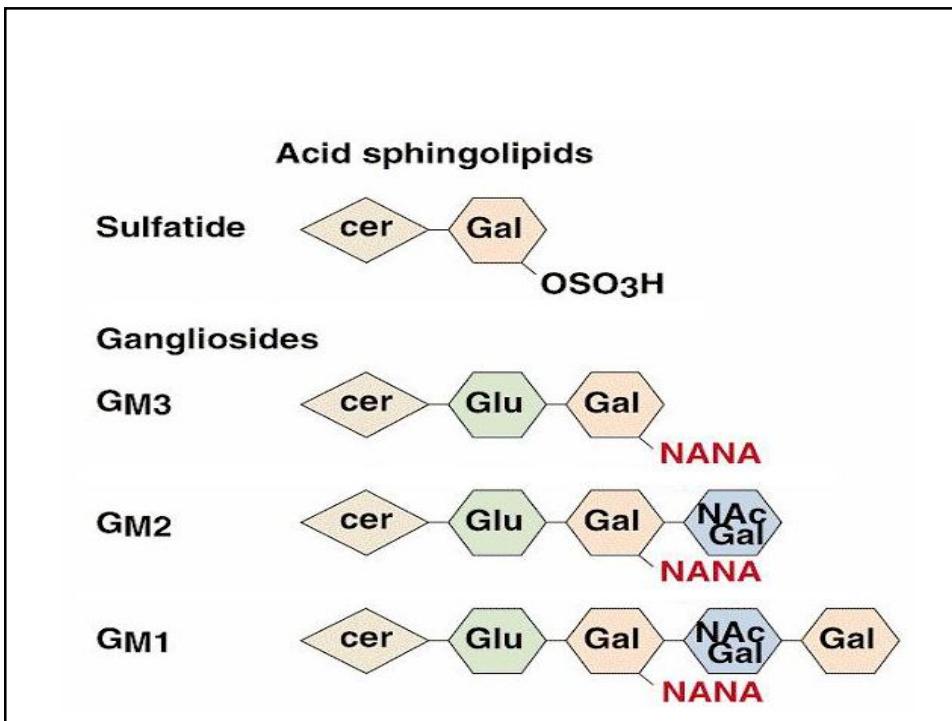
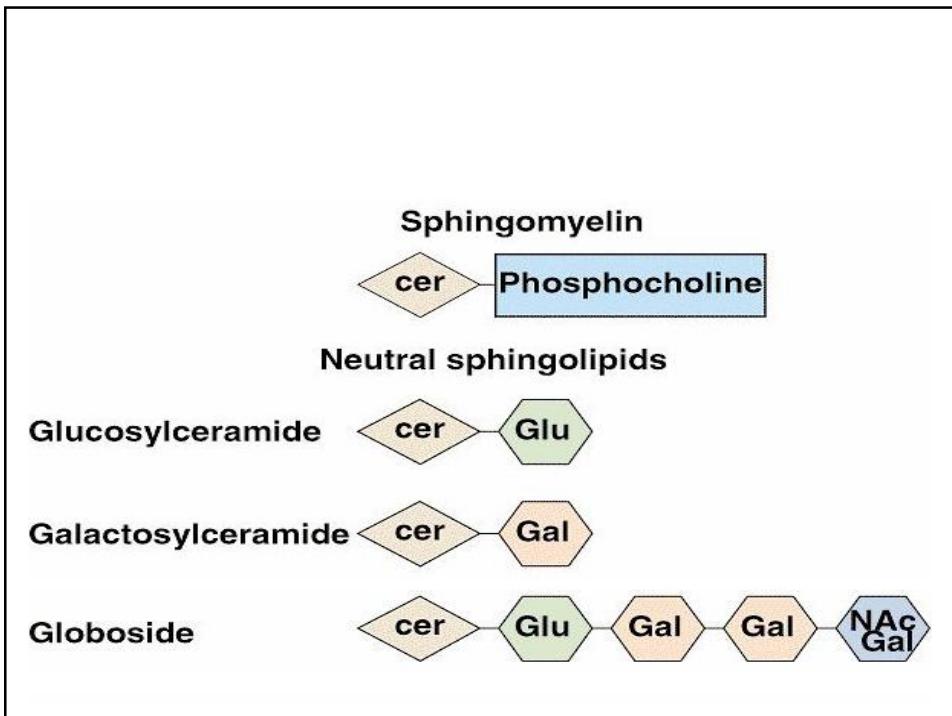
-Oligosaccharide => Globoside

-Oligosaccharide with NANA => Gangliosides

## N-Acetylneuraminic Acid (NANA)

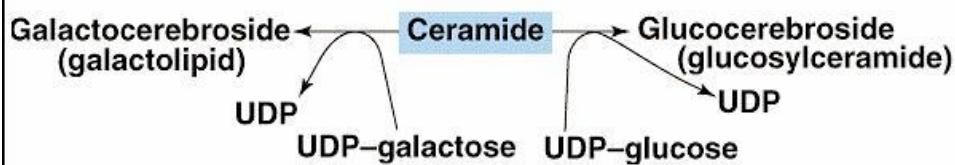


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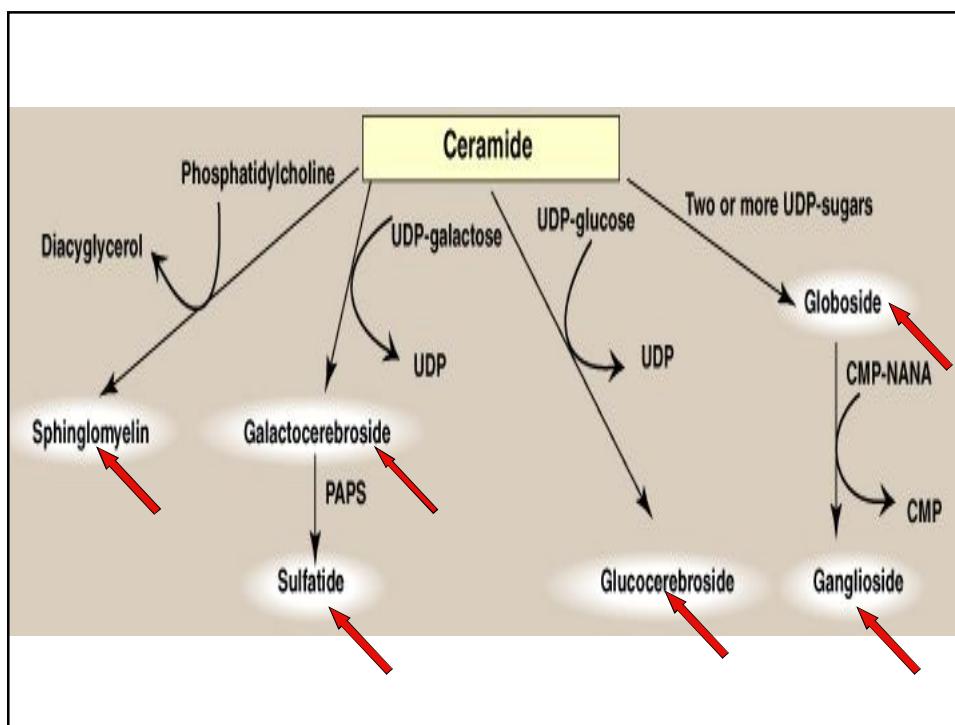
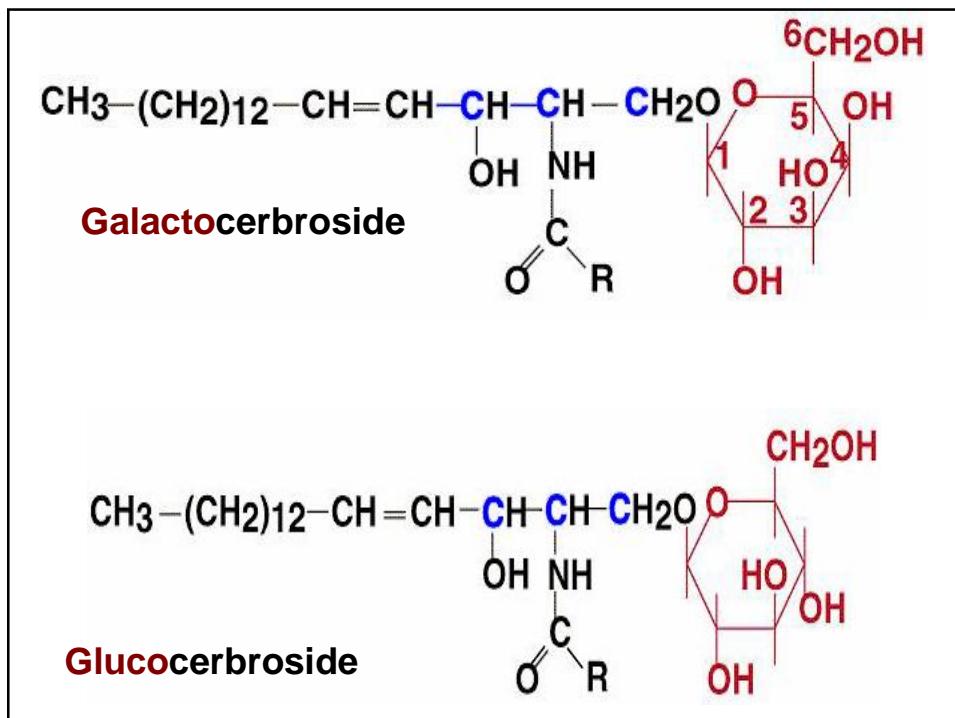


## Activated Donors in Glycolipids Synthesis

- UDP-Glucose
- UDP-Galactose
- UDP-N-Acetylgalactosamine
- CMP- N-Acetylneurameric Acid

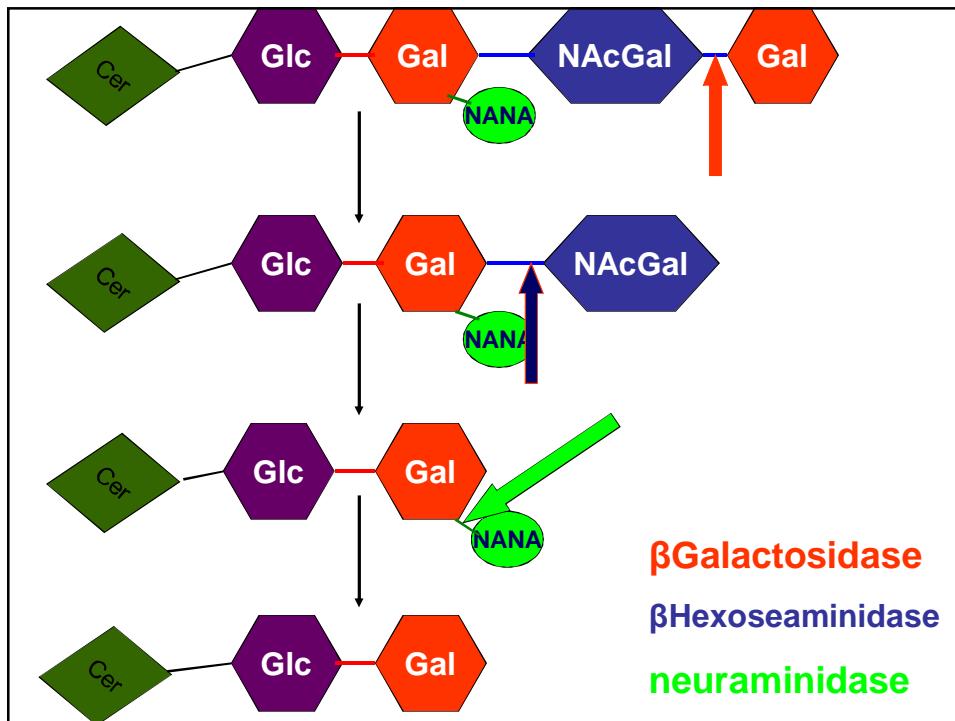


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## Degradation of Sphingolipids

- Hydrolytic Enzymes, Specific for the Sugar
- - $\alpha$  Galactosidase
  - $\beta$  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.

