



Textbook of BIOCHEMISTRY for Medical Students

TENTH EDITION

Vasudevan



Lipids may be **defined as** compounds which are relatively insoluble in water, but freely soluble in nonpolar organic solvents like benzene, chloroform, ether, hot alcohol, acetone, etc.



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Functions of Lipids



- 1. Storage form of energy (triglycerides)
- 2. Structural components of biomembranes
- 3. Metabolic regulators (steroid hormones)
- 4. Act as surfactants, detergents and emulsifying agents (amphipathic lipids)
- 5. Act as electric insulators in neurons
- 6. Provide insulation against changes in external temperature (subcutaneous fat)
- 7. Give shape and contour to the body
- 8. Protect internal organs (pads of fat)
- 9. Help in absorption of fat soluble vitamins (A, D, E and K)

Clinical Applications of Lipids



- 1. Excessive fat deposits cause obesity. Truncal obesity is a risk factor for heart attack.
- 2. Abnormality in cholesterol and lipoprotein metabolism leads to atherosclerosis and cardiovascular diseases.
- 3. In diabetes mellitus, the metabolisms of fatty acids and lipoproteins are deranged, leading to ketosis and dyslipidemia.



Classification of Lipids

I. Simple lipids

- a. Triacylglycerol or triglycerides or neutral fat
- b. Waxes
- **II. Compound lipids**
- A. Phospholipids, containing phosphoric acid
- 1. Nitrogen containing glycerophosphatides:
- i. Lecithin (phosphatidylcholine)
- ii. Cephalin (phosphatidylethanolamine)
- iii. Phosphatidylserine
- 2. Non-nitrogen glycerophosphatides
- i. Phosphatidylinositol
- ii. Phosphatidylglycerol
- iii. Diphosphatidylglycerol (cardiolipin)

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Classification of Lipids, Continued

- 4. Phosphosphingosides, with sphingosine Sphingomyelin
- B. Non-phosphorylated lipids
- 1. Glycosphingolipids (carbohydrate)
- i. Cerebrosides (ceramide monohexosides)
- ii. Globosides (ceramide oligosaccharides)
- iii. Gangliosides (ceramide + oligosaccharides + N-acetylneuraminic acid)
- 2. Sulfolipids or sulfatides
- i. Sulfated cerebrosides
- ii. Sulfated globosides
- iii. Sulfated gangliosides
- **III. Derived lipids**

Fatty acids, steroids, prostaglandins, leukotrienes, terpenes, dolichols, etc.

IV. Lipids complexed to other compounds

Proteolipids and lipoproteins

I. <u>Simple Lipids</u>

- a) Triacyl glycerol or Triglycerides or neutral fat
- b) Waxes
- II. <u>Compound Lipids</u>
- III. <u>Derived Lipids</u>

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- fatty acids, steroids, prostaglandins
- **IV. Lipids Complexed to Other**

Proteolipids and lipoproteins

Compounds

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Fatty Acids



Derived lipids.

They are generally found in ester linkage in different classes of lipids. In the human body free fatty acids are formed only during metabolism.

Fatty acids are **aliphatic carboxylic acids** and have the general structural formula, R - CO - OH, COOH (carboxylic group)





- **1. Depending on total number of carbon atoms:**
- **a.** Even chain: They have carbon atoms 2, 4, 6 and similar series.
- **b.** Odd chain: They have carbon atoms 3, 5, 7, etc. They are present in milk.
- 2. Depending on length of hydrocarbon chain:
- a. Short chain with 2 to 6 carbon atoms
- b. Medium chain with 8 to 14 carbon atoms
- c. Long chain with 16 and above, usually up to 24 carbon atoms
- d. Very long chain fatty acids (more than 24 C).
- 3. Depending on nature of hydrocarbon chain:
- a. Saturated fatty acids
- b. Unsaturated fatty acids
 - i) Monounsaturated (monoenoic)
 - ii) Polyunsaturated (polyenoic)

Saturated Fatty Acids



Human body fat contains 50% oleic acid, 25% palmitic acid 10% linoleic and 5% stearic acid.

The carbon atoms of fatty acids are numbered as C1, C2 etc starting from the COOH group. Or, starting from the methyl end, the carbon atoms may be numbered as omega (ω)-1,2,3, etc.

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6 5 4 3 2 1 CH3 CH2 CH2 CH2 CH2 CH2 COOH ω1 ω2 ω3 ω4 ω5



Common	No	Chemical nature	Occurrence				
name	carbo	n 🚁 😡					
	atoms						
Even chain, Saturated fatty acids							
Acetic	2	Saturated; small chain	Vinegar				
Butyric	4	do Diagnostic testing for COVID -19 included	Butter				
Caproic	6	do	Butter				
Lauric	12	do	Coconut oil				
Palmitic	16	Saturated; long chain	Body fat				
Stearic	18	do DM Vasudevan	do				
Odd-chain fatty acids							
Propionic	3	Saturated; Odd chain	Metabolism				



Unsaturated fatty acids

Common name	No. of carbon atoms	Chemical nature	Occurrence
Palmitoleic	16	Monounsaturated (w7)	Body fat
Oleic	18	do (w9)	do
Erucic	22	do (w9)	Mustard oil
Nervonic	24	do (w9)	Brain lipids
Linoleic	18	2 double bonds (w6)	Vegetable oils
Linolenic	18	3 double bonds (w3)	do
Arachidonic	20	4 double bonds (w6)	Vegetable oils







They exhibit geometrical isomerism at the double bonds. All the naturally occurring fatty acids have the **cis configuration**.

 $\begin{array}{c} \mathsf{CH}_3 - (\mathsf{CH}_2)_7 - \mathsf{CH} \\ \| \\ \mathsf{HOOC} - (\mathsf{CH}_2)_7 - \mathsf{CH} \\ \mathsf{HOOC} - (\mathsf{CH}_2)_7 - \mathsf{CH} \\ \end{array} \\ \begin{array}{c} \mathsf{HOOC} - (\mathsf{CH}_2)_7 - \mathsf{CH} \\ \mathsf{HOOC} - (\mathsf{CH}_2)_7 - \mathsf{CH} \\ \end{array} \\ \end{array}$

Cis-form (Oleic acid)

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Clinical Significance of PUFA



- 1. Example, Linoleic and Linolenic acids
- 2. They are called essential fatty acids, because they cannot be synthesized by the body and have to be supplied in the diet.
- 3. Unsaturated fatty acids are also designated as ω 3 (omega 3) family—Linolenic acid ω 6 family—Linoleic and Arachidonic acids ω 9 family—Oleic acid
- 4. Arachidonic acid is the precursor of prostaglandins. Arachidonic acid can be synthesized in the body.
- 5. The pentanoic acid present in fish oils is of great nutritional importance (ω 3 unsaturated fatty acid).
- 6. Eicosanoids (eicosa = twenty) are derived from 20 C arachidonic acid. They are polyenoic fatty acids. They are precursors of prostaglandins.

Trans Fatty Acids (TFA)



- TFA are present in dairy products and in hydrogenated edible oils.
- They are generally considered to be injurious to health.
- However, TFA are used in food industry as they increase the shelf life of the fried food.
- Oils containing PUFA also have high content of TFA.
- Fast food preparations have a high TFA content.
- **Trans fatty acids** adversely affect composition of blood lipids and lipoproteins, systemic inflammation, endothelial dysfunction, insulin resistance, diabetes and adiposity.



Hydrogenation

Unsaturated fatty acids may be converted to the corresponding saturated fatty acids by hydrogenation of the double bond.

 $(+)2H \qquad (+)2H \qquad (+)2H$ Linolenic — \rightarrow Linoleic — \rightarrow Oleic — \rightarrow Stearic

Hydrogenation of oils can lead to solidification and saturation, e.g. Vanaspathi.



Halogenation



When treated with iodine, the unsaturated fatty acids can take up two halogen atoms, at each double bond.

For example,

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Oleic acid + $I_2 \rightarrow$ Di-iodo oleic acid

The number of halogen atoms taken up will depend on the number of double bonds and is an index of the degree of unsaturation.



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Physical Characteristics



The short and medium chain fatty acids are liquids, whereas long chain fatty acids are solids at 25oC.

Melting and boiling points increase, with increase in chain length.

The unsaturated fatty acids have lower melting point compared to saturated fatty acids with the same chain length.



Melting Point



- The short and medium chain fatty acids are liquids, whereas long chain fatty acids are solids at 25°C.
- The solubility in water decreases, while melting and boiling points increase with increase in chain length.
- The unsaturated fatty acids have lower melting point compared to saturated fatty acids with the same chain length.
- For example, stearic acid (C18 fatty acid, no double bond) has the melting point 69°C, oleic acid (C18, 1 double bond) has 13°C; linoleic acid (C18, 2 double bonds) has -5°C and linolenic (C18, 3 double bonds) has -10°C.

Salt Formation



Saturated and unsaturated fatty acids form salts with alkali.

$CH3-COOH + NaOH \rightarrow CH3-COONa + H2O$

Sodium and potassium salts of long chain fatty acids are called **soaps**.

Calcium and magnesium soaps are insoluble.



Ester Formation



 $Glycerol + fatty acid \rightarrow Mono acyl glycerol$ $Monoglyceride + fatty acid \rightarrow Di acyl glycerol$ $Diglyceride + fatty acid \rightarrow Triglyceride$ or tri acyl glycerolor neutral fat

Nomenclature of Carbon Atoms



- As per International Union of Biochemistry (IUB) the correct designations are monoacylglycerol (MAG), diacylglycerol (DAG) and triacylglycerol (TAG).
- But the old terminology of monoglyceride, diglyceride and triglyceride are still popular, especially among clinical laboratory workers.
- The carbon atoms of glycerol are designated as a, b and a' or as 1, 2, 3, where R represents the side chain of fatty acids.
- Enzymes can distinguish between 1st and 3rd carbon atoms.

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Composition of Oils and Fats



Name	Saturated fatty acids(%)	Mono- unsaturated fatty acids(%)	PUFA (%)		
Coconut oil	(*)86	12	2		
Groundnut oil	18	46	36		
Gingelly oil (Til oil)	13	50	37		
Palm oil	42	52	6		
Corn oil	13	25	62		
Cotton Seed oil	26	19	55		
Mustard oil (rapeseed)	34(**)	48	18		
Safflower oil (Kardi)	9	12	79		
Sunflower oil	12	24	64		
Butter	75	20	5		
Ox (Tallow)	53	42	5		
Pig (Lard)	42	46	12		
Fish oil	30	13	57		
(*) these saturated fatty acids are medium chain fatty acids.					
(**) contains arusis said 22 C 1 double hand)					

**) contains erucic acid, 22 C, 1 double bond)

Physical Properties of Triglycerides



Oils are liquids at 20oC; they are triglycerides with unsaturated fatty acids or short chain triglycerides. Oils are generally of plant origin.

Fats are solids at room temperature and contain mainly saturated long chain fatty acids. Fats are mainly of animal origin.

Fats containing medium chain triglycerides or unsaturated fatty acids are soft fats, e.g. butter, coconut oil. Coconut oil contains mainly medium chain TAG, e.g. Lauric and Myristic acids.



Mixed Triglycerides



- Naturally occurring fats and oils are mixtures of triglycerides.
- If all the three hydroxyl groups of the glycerol are esterified to the same fatty acid, a simple triacylglycerol is formed, e.g. Tripalmitin, Triolein, etc.
- A mixed triacylglycerol is formed, when different fatty acids are esterified to the hydroxyl groups of glycerol.
- Generally, two hydroxyl groups are esterified to similar fatty acid and the third with a different one, e.g. palmitic acid in 1 and 3 positions, while oleic acid is in the 2nd position.
- When a PUFA is present, it is commonly esterified to the 2nd or b-carbon atom.

Physical Properties of Triacylglycerols

THE REAL

- They are hydrophobic and insoluble in water.
- Oils are liquids at 20°C; they are triacylglycerols, which contain a higher proportion of unsaturated fatty acids or short chain triglycerides.
- Oils are generally of plant origin.





- Fats are solids at room temperature and contain mainly saturated long chain fatty acids.
- Fats are mainly of animal origin.
- When the constituent fatty acids have a higher chain length and are predominantly saturated, 'hard fat' is formed, e.g. pig fat.
- Fats containing medium chain triacylglycerols or unsaturated fatty acids are soft fats, e.g. butter, coconut oil. Coconut oil contains mainly medium chain TAG, e.g. lauric and myristic acids.

Storage of Energy as Fat



The triacyl glycerides are the storage form of lipids in the **adipose tissue**.

When stored as TAG, water molecules are repelled and space requirement is minimal.

Excess fat in the body leads to obesity.

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Hydrolysis of Triglycerides



Triacyl glycerol is **sequentially hydrolysed** to diacyl glycerol and mono acyl glycerol and finally glycerol plus 3 fatty acids.



Saponification



When triglycerides are hydrolysed by alkali, the process is known as saponification. The products are glycerol and soaps. Saponification number is defined as the number of milligrams of potassium hydroxide required to saponify one gram of fat. It is an indication of the molecular weight of the fat, and is inversely proportional to it.



Iodine Number of a Fat



It is defined as the number of grams of iodine taken up by 100 grams of fat.

It is an index of the degree of unsaturation and is directly proportional to the content of unsaturated fatty acids. Higher the iodine number, higher is the degree of unsaturation.

e.g., iodine number of butter is 28, and that of sunflower oil is 130.



Rancidity of Fat



- Fats and oils have a tendency to become rancid.
- The term rancidity refers to the appearance of an unpleasant smell and taste for fats and oils.
- Hydrolytic rancidity is due to partial hydrolysis of the triacylglycerol molecules due to traces of hydrolytic enzymes present in naturally occurring fats and oils.





- Oxidative rancidity is the result of partial oxidation of unsaturated fatty acids with resultant formation of epoxides and peroxides of small molecular weight fatty acids by peroxides and free radicals.
- The same process, if it occurs in vivo will affect the integrity of biomembranes, leading to cell death.





- Many natural fats and oils may contain antioxidants (e.g. vitamin E), which prevent the occurrence of oxidative rancidity.
- PUFA are more easily oxidized; so vegetable oils with a high content of PUFA are usually preserved with addition of antioxidants.
- Repeated heating of oils would lead to the formation and polymerization of cyclic hydrocarbons.
- These will impart an unpleasant taste and color to the oil.
- Coconut oil having medium chain saturated fatty acids will withstand such polymerization.

Waxes



- They form the secretions of insects, leaves and fruits of plants, e.g. Lanolin or wool fat, beeswax, whale sperm oil, etc.
- They are esters of higher fatty acids with higher monohydroxy aliphatic alcohols and so have very long straight chains of 60–100 carbon atoms.
- They are used as the base for the preparation of cosmetics, ointments, polishes, lubricants and candles.



Compound Lipids



A) Phospholipids, with phosphoric acid. 1. Nitrogen containing glycerophosphatides i) Lecithin (phosphatidyl choline) ii) Cephalin iii) Phosphatidyl serine 2. Non-nitrogen glycerophosphatides Phosphatidyl inositol **i**) ii) Phosphatidyl glycerol iii) Diphosphatidyl glycerol 3. Plasmalogens, having long chain alcohol **i**) Choline plasmalogen ii) Ethanolamine plasmalogen 4. Phospho sphingosides, with sphingosine Sphingomyelin

Compound Lipids Continued

THE REAL

- A)Phospholipids, with phosphoric acid
- **B) Non-phosphorylated lipids**
 - 1. Glycosphingolipids (carbohydrate)
 - i) Cerebrosides (ceramide monohexosides)
 - ii) Globosides (ceramide oligosaccharides)
 - iii) Gangliosides (having N-acetyl neuraminic acid)
- 2. Sulpholipids or sulfatides
 - i) Sulphated cerebrosides
 - ii) Sulphated globosides
 - iii) Sulphated gangliosides

Phospholipids



They contain glycerol, fatty acids and a nitrogenous base.

A) Phosphatidates; derivatives of phosphatidic acid.



Phosphatidicacidhasoneglycerolandtwofattyacidresidues.is3rdhydroxylgroupisesterified to a phosphoric acid



1. Amphipathic Nature

They have both hydrophobic and hydrophilic portion in their molecule.

2. Micellar Formation

Their hydrophobic parts keep away from water, forming molecular aggregates called micelle. help in digestion and absorption of lipids

3. Biomembranes

The molecules align themselves to form monolayers with the polar heads pointing in one direction and the nonpolar tails in the opposite direction

Biomembranes



- The molecules align themselves to form monolayers with the polar heads pointing in one direction and the nonpolar tails in the opposite direction.
- Only fatty acids with more than 6 carbon atoms form monolayers.
- This explains their role as components of biomembranes.
- The self-assembly of phospholipids into bilayers is driven by hydrophobic interaction.
- They also act as detergents and emulsifying agents.
- In vivo, they act as pulmonary surfactants.







4. Liposomes

Liposomes are microscopic spherical vesicles. The phospholipids arrange themselves to form a bilayer membrane which encloses some of the water in a phospholipid sphere.

Drugs, proteins, enzymes, etc. may be encapsulated by the liposomes which could act as carriers for these substances to target organs.

Liposomes have important applications in cancer chemotherapy, antimicrobial therapy, gene therapy, vaccines and diagnostic imaging.

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Phospholipids form micelles and liposomes

Aquasomes



- They are one of the recently developed delivery systems that act as carrier systems for delivery of peptide based pharmaceuticals.
- These are nanoparticulate carrier systems.
- They comprise the central solid nanocrystalline core coated with polyhydroxy oligomers onto which biochemically active molecules are adsorbed.
- The solid core provides the structural stability.
- The carbohydrate coating stabilizes the biochemically active molecules.
- The delivery system has been successfully utilized for the delivery of insulin, hemoglobin and various antigens.
- Oral delivery of enzymes like serratiopeptidase has also been achieved.

Amphipathic Nature



- Phospholipids in general are amphipathic, particularly Lecithin.
- They have both hydrophobic and hydrophilic portion in their molecule.
- The glycerol along with the phosphoric acid and choline constitute the polar 'head' of a phospholipid molecule, whereas the hydrocarbon chains of the fatty acids represent the non-polar 'tail'.



Phosphatidyl Choline or Lecithin



Nitrogen containing phospholipid. It contains the glycerol group. The alpha and beta positions are esterified with fatty acids. (betacarbon with a **PUFA** molecule) Phosphoric acid is added to 3 position Phosphate group is esterified to the quaternary nitrogen base, Choline

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Action of Phospholipases



Lecithin. R1 and R2 are fatty acids. Red rectangle depicts glycerol group. The blue rectangle is choline which shows polar or hydrophilic property.

- 1 = Site of action of phospholipase A1;
- 2 = Site of action of phospholipase A2;
- 3 = Site of action phospholipase C;
- 4 = Site of action of phospholipase D.





Pulmonary Surfactants



Pulmonary surfactant is a surface-active lipoprotein (phospholipoprotein) formed by alveolar cells. The molecule is seen at the air-water interface of alveoli. The hydrophilic head dips in water and the hydrophobic tail faces towards the air, so that the surface tension is reduced.

Pulmonary surfactant reduces surface tension at the air-liquid interface of the alveolus, thus preventing its collapse during endexhalation. Surfactant also participates in innate host defense against inhaled pathogens.



Composition of Surfactant



Dipalmitoylphosphatidylcholine (DPPC), 40%; other phospholipids, 40%; surfactant proteins(SP-A,SP-B,SP-C, SP-D) 5%; cholesterol 5%.

The main lipid component of surfactant is DPPC. It has two 16carbon saturated fatty acids, a phosphate group and a choline group attached. The DPPC is the strongest surfactant molecule in the pulmonary surfactant mixture.

Phosphatidylcholine forms 85% of the lipid in surfactant. Phosphatidyl glycerol forms about 10% of the lipids in the surfactant.



Respiratory distress syndrome (RDS): Morbidity in preterm neonates. Patients present shortly after birth with apnea, cyanosis, grunting, inspiratory stridor, nasal flaring, poor feeding, and tachypnea.

Radiological findings include a diffuse "ground glass" appearance (resulting from alveolar atelectasis).

The preterm infant who has RDS has low amounts of surfactant. It is due to a defect in the biosynthesis of dipalmitoyl lecithin (DPL), the main pulmonary surfactant.

Acute Respiratory Distress Syndrome (ARDS)



It is a syndrome of acute pulmonary inflammation. It is characterized by sudden onset, impaired gas exchange, and pulmonary edema. Infection is the most common cause of development of ARDS in children. The lungs appear particularly vulnerable in the first year of life.

There is an increase in the permeability of the alveolar capillary arrier as a result of injury to the endothelium.

Damage to the alveolar cells leads to an influx of edema fluid into the alveoli, as well as decreased fluid clearance from the alveolar space.

Meconium Aspiration Syndrome



Meconium staining of the amniotic fluid or fetus is an indication of fetal distress.

In the presence of fetal distress, gasping may be seen in utero.

This leads to aspiration of amniotic fluid along with meconium, into the large airways.

It is characterized by airway obstruction, pneumonitis, pulmonary hypertension, acidosis and hypoxemia.

Meconium destroys surfactant and decreases its surface adsorption rate.

Neonatal Hyaline Membrane Disease



Waxy-appearing layers of hyaline membrane are seen lining the alveoli of the lung. Hyaline membranes are composed of fibrin, cellular debris, red blood cells, neutrophils and macrophages. As a result, the blood passing through the lungs is unable to pick up oxygen and unload carbon dioxide. The condition is also called neonatal respiratory distress syndrome (RDS), or respiratory distress syndrome of newborns. It is seen in premature infants and is caused by insufficiency of pulmonary surfactant production. It begins shortly after birth and is manifested by fast breathing, increased heart rate, blue discoloration of the skin and apnea. As a treatment, oxygen is given with continuous positive airway pressure (CPAP). Intravenous fluids are given to stabilize blood pressure.



In pregnancies of greater than 30 weeks, the fetal lung maturity may be tested by sampling the amount of surfactant in the amniotic fluid. Several tests are available that correlate with the production of surfactants. These include the lecithin-sphingomyelin ratio ("L/S ratio") and the presence of phosphatidylglycerol. If the L/S ratio is less than 2:1, the fetal lungs may be surfactant deficient. The presence of PG usually indicates fetal lung maturity.

Phosphatidyl Ethanolamine or Cephalin



The nitrogen base ethanolamine is present (instead of choline in lecithin)

Cephalin is also found in biomembranes and possesses amphipathic properties.





Phosphatidic acid is esterified to inositol.

Phosphatidyl inositol bisphosphate or **PIP2** is present in biomembranes. This plays a role in hormone action.



Plasmalogens



Phospholipids with an aliphatic long chain unsaturated alcohol in the first position of glycerol. The second OH group is esterified to a fatty acid.

The phosphoric acid is attached to choline or ethanolamine.

Plasmalogens are found in biomembranes in brain and muscle.

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It is formed by esterification of phosphatidic acid to glycerol.

When two molecules of phosphatidic acid are linked with a molecule of glycerol, diphosphatidyl glycerol or **cardiolipin** is formed.

This phospholipid is found in myocardium.





All sphingolipids have the long aliphatic amino alcohol sphingosine

It is attached to a fatty acid in amide linkage to form a **ceramide**. The fatty acid has a chain length varying from C18 to C24.









They contain **phosphoric acid** group.

Sphingomyelins are the only sphingolipid that contain phosphate and have no sugar moiety.





Because of its amphipathic nature, sphingomyelin can act as an emulsifying agent and detergent.

The relative proportion of lecithin and sphingomyelin is important in biological fluids like bile, amniotic fluid, etc.

Sphingomyelin combined with fatty acid is called **ceramide**, which is a component of glycosphingolipids.



APL Antibody Syndrome



Antiphospholipid antibodies have been found to be present in blood of patients with autoimmune diseases and of patients with thrombotic episodes. In pregnancy, the presence of these antibodies can cause complications like miscarriage, eclampsia and preterm labor. Here the antibodies are developed against membrane phospholipid, **cardiolipin**.

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Non-phosphorylated Lipids



<u>Glycosphingolipids</u> (Glycolipids)

They are seen widely in nerves.



This group of lipids do not contain phosphoric acid; instead they contain carbohydrates and ceramide.

Ceramide + Glucose \rightarrow Gluco cerebroside

Ceramide + Galactose \rightarrow Galacto cerebroside

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Globosides (Ceramide Oligosaccharides)



They contain two or more hexoses, attached to a ceramide molecule

Ceramide + Galactose + Glucose \rightarrow Lactosyl ceramide

It is a component of erythrocyte membrane.

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 Updated Long & Short Qs and Esser.

New MCOs and Case studies

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Gangliosides



They are formed when ceramide oligo-saccharides have one or more molecules of NANA (N-acetyl neuraminic acid) (sialic acid) attached to them.

Ceramide—Glucose—galactose— NANA;

this is designated as ganglisoside M3 (GM3).





Sulfate groups are attached to ceramide polysaccharides.

Important components of membranes of nervous tissue.

Failure of degradation of these compounds results in accumulation of these complex lipids in CNS. This group of inborn errors is known as lipid storage diseases.

