وَقُل رَّبِّ زِدْنِي عِلْمًا

النادي الطبي

RESPIRATORY SYSTEM

HAYAT BATCH

SUBJECT : Biochemistry

LEC NO. : 1

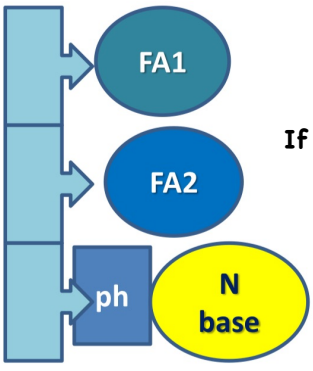
DONE BY : Esra'a Khaled

Respiratory System

في هاي المحاضرة رح نحكي عن 3 أمراض :

- Respiratory distress syndrome (RDS)
- α1-Antitrypsin (α1- antiproteinase) deficiency
- Cystic fibrosis

* بدايةً رح نحكي عن ال **phospholipids** الي بدخل في تركيب ال **lung surfactant** و الي بكون مش موجود في حالة ال RDS



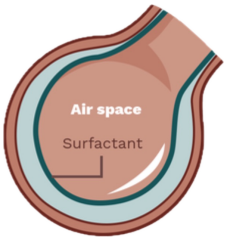
If the alcohol is glycerol → Glycerophospholipids = phosphoglycerides

* Phosphatidic acid : This is the simplest phosphoglyceride
* Phosphatidylcholine (lecithin) : The phosphate group of phosphatidic acid was esterified with choline

* Dipalmitoyl phosphatidylcholine (DPPC) : made and secreted by granular pneumocytes (Type II pneumocytes) , is the major lipid component of lung surfactant , which is synthesized shortly before parturition in full-term infants

* Surfactant serves to :

1. decrease the surface tension of this fluid layer
2. reducing the pressure needed to reinflate alveoli
3. preventing alveolar collapse (atelectasis)



* Respiratory distress syndrome (RDS) in pre-term infants is associated with insufficient surfactant production
* Lung maturity of the fetus can be gauged by determining the ratio of DPPC to sphingomyelin , L (lecithin) / S ratio , in amniotic fluid , ⩾ 2 is evidence of maturity
* Lung maturation can be accelerated by giving the mother glucocorticoids shortly before delivery
* Insulin inhibits pneumocytes and decrease lung surfactant , sometimes diabetic mother delivers a baby with RDS
* Also RDS can occur in adults whose surfactant-producing pneumocytes have been damaged or destroyed
  For example :
  adverse side effect of immunosuppressive medication or chemotherapeutic drug use

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Respiratory System

* هلاً نيجي للمرض التاني : α1-Antitrypsin (α1- antiproteinase) deficiency

• α1- antiproteinase, is produced by the hepatocytes and macrophages .
It forms complexes with plasma serine proteases (trypsin and elastase) , inhibiting their activity .

* excess elastase activity is checked by α1-antiproteinase , Thus deficiency of this protein causes damage to lung tissue , leading to emphysema.

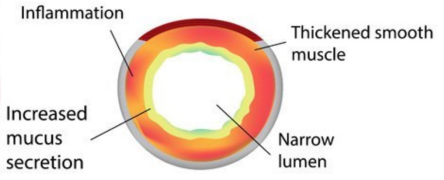
* آخر مرض : Cystic fibrosis

* It is an inherited autosomal recessive disorder , characterized by :

1. chronic bacterial infections
2. Fat maldigestion
3. Infertility in male
4. Elevated levels of chloride in a sweat > 60 moo/L

- The cystic fibrosis gene known as CFTR occurs on chromosome 7 and encode a protein of 1480 amino acids, named cystic fibrosis transmembrane regulator (CFTR), a cyclic AMP-regulated **CL^- channel.**

* Any abnormality of membrane CL- permeability is believed to result in the increased viscosity of many bodily secretions .

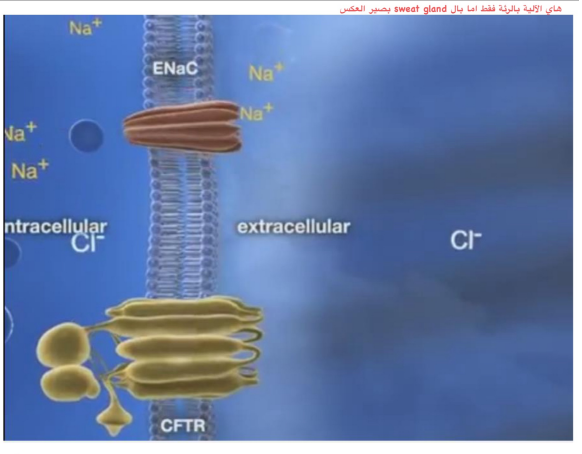


* deletion of 3 bases resulting in loss of residue 508 , a phenylalanine so the mutant allele is three bases shorter than the normal allele .

* it is possible to distinguish them from each other by the size of the PCR products obtained by amplifying that portion of the DNA .

CFTR not only allows chloride ions to be drawn from the cell and into the ASL, but it also regulates another channel called ENaC (**Epithelial Sodium Channel)**, which allows sodium ions to leave the ASL and enter the respiratory epithelium.

Defective CFTR results in decreased secretion of chloride and increased reabsorption of sodium and water across epithelial cells. The resultant reduced height of epithelial lining fluid and decreased hydration of mucus results in mucus that is stickier to bacteria, which promotes infection and inflammation.



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