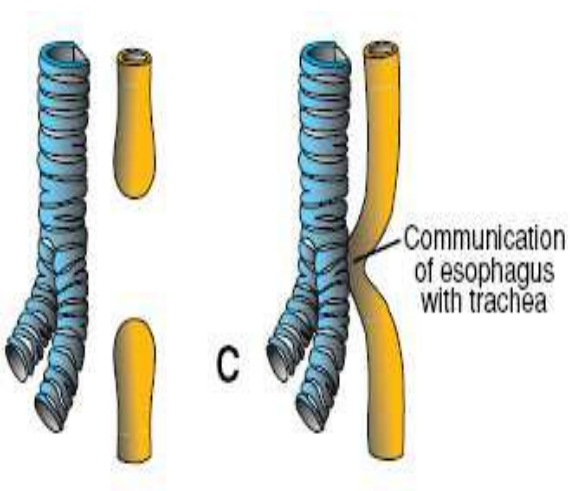
Respiratory system
Embryology (2)
Anomalies of development of RT

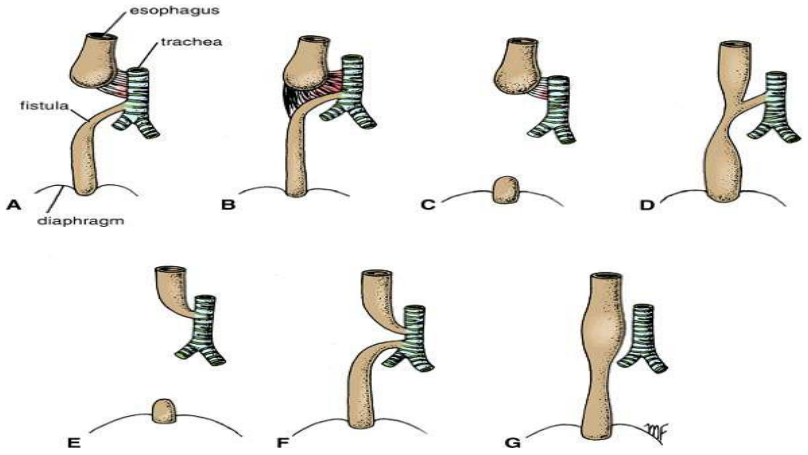
-In the previous lecture we talked about development of the nose, palate (primary and secondary) and its clinical application (cleft lip and palate), and respiratory diverticulum (respiratory bud)

Anomalies of trachea and esophagus:
A) TEF (tracheoesophageal fistula)
- We have different forms of TEF:
1) Proximal esophageal atresia accompanied by distal fistula:
- occurs 1/3000 births
-account for 90% of TEF (tracheoesophageal fistula)
-predominant in male infants

2) Isolated esophageal atresia and H-type TEF :(as in the picture below to the left)
- Account for 4% of TEF
- It appears as double atresia or it appear as H-shaped

3) Other variations account for 1% of TEF (as in the picture below to the right)





-some clinical points concerning esophageal atresia
1) Infant suffers from cough and chokes which occur due to excessive saliva and intake which accumulate as a result of blockade of its way because of proximal esophageal atresia
2) Polyhydramnios: One of the complications that occur because of the blockade (due to atresia) where all the amniotic fluid returns to the sac leading to polyhyramnios (opposite to oligohydramnios)
3) Gastric contents will be filled with air so it will be distended (especially during infant crying)
4) Nothing can reach to the gastric content because of the obstruction
5) Fistula can lead to pneumonitis (pneumonia /infection of the lung)

-other anomalies associated with esophageal anomalies:
1) Cardiac defects:

- Most common anomaly associated with TEF
-present in 33% of patients with TEF
- Anomalies that might affect the heart include: ventricular septal defect, atrial septal defect, and fallots
2) Vertebral anomalies
3) Anal atresia (closure of anal canal)
4) Limb defects

B) Tracheal atresia and stenosis

-It might occur with esophageal anomalies (TEF) as we mentioned previously
-It might be due to obstruction of trachea by tissue:
Occur due to defect in separation of trachea and esophagus so you find that there is a tissue filling the trachea (this separation might not be in the proper site "more toward the esophagus" so trachea is stenosed –incomplete tracheal atresia-)

Anomalies of the larynx:

-we talked about development of the larynx, and its cartilage which has mesenchymal origin (derived from the mesoderm) ,and we talked about opening of the larynx and how it changes from slit like to T-shaped and then it assumes its final shape

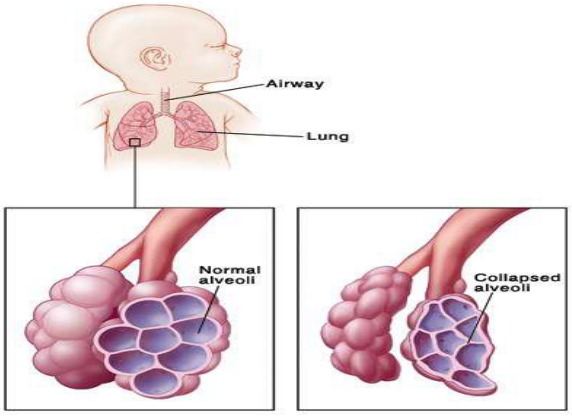
-Anomalies of the larynx include **laryngeal atresia:**

– It means that the larynx is blocked (blind)
- It's a rare anomaly
- It affects upper parts of larynx so it's called congenital highway obstruction syndrome
- Organs distal to the atresia will be enlarged particularly the lungs to compensate for the reduced amount of air that might pass through the atresia
- Diaphragm is either flattened or inverted and this is the cause of obstruction
- One of the complications is fetal ascites (accumulation of fluid in abdominal cavity)
- You might find serous fluid around the lung
-Ultrasonography is one of the techniques used in diagnosis

Anomalies of the lung:

-One of the common anomalies is RDS:

-notice on the picture below that normal alveoli are completely distended; also notice the collapsed alveoli seen in RDS "in this syndrome expansion of the wall of alveoli isn't complete"
-account about 30% of all neonatal disease



*The partially collapsed alveoli contain a fluid with a high protein content, many hyaline membranes and lamellar bodies; and that's why RDS is known as **Hyaline membrane disease** (it forms a membrane on the alveolar wall from inside). These collapsed alveoli are mostly derived from the surfactant layer.

*So hyaline membrane disease accounts for **20%** of deaths among newborns

Intrauterine Asphyxia might occur if there're changes in type II cells; meaning that there's NO secretion of surfactants.

-Intrauterine Asphyxia means that the baby will die in the uterus because there's NO secretion.

*The amniotic fluid (will be discussed later in this sheet) stimulates the development of alveoli and a little bit of respiration might take place inside, although it doesn't occur, leading to the inflation. If this does NOT occur, Asphyxia will happen.

Treatment:

*Recent development of artificial surfactant and treatment of premature babies with **corticosteroids (betamethasone)** to stimulate surfactant production (affecting type II cells) have reduced the mortality associated with RDS.

*Also they found that **Thyroxine**, which is secreted from the thyroid gland, is the most important stimulator for surfactants formation (so we could give Thyroxine as a Tx)

*Giving the injection to babies as young as 5.5 months of gestation allows survival of these babies.

Other anomalies:

Abnormalities of Lung and bronchial tree (e.g. blind- ending trachea or absence of lungs (lung agenesis) which could be either one lung*unilateral* or bilateral and, even these cases are rare, there'll be no inflation)

Abnormal divisions of the bronchial tree, some result in supernumerary lobules (excess # of the bronchopulmonary segments or the lobules of the lung (more division))

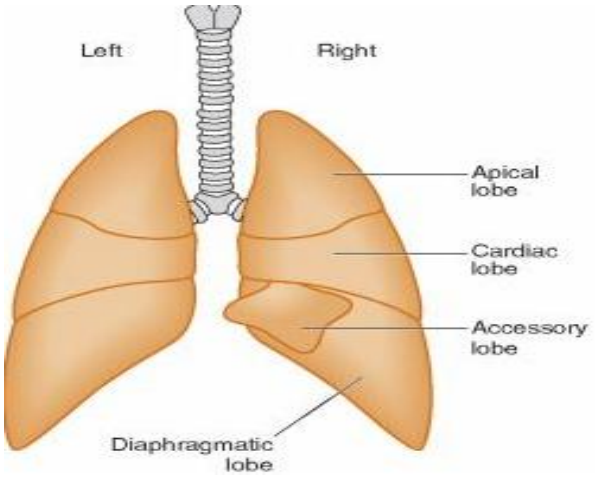
*If the # is increased, does it affect the baby? Because there's an increasing, it will NOT affect the baby but its effect will be negative if the doctor used the bronchoscope and he thought that there's a normal distribution of bronchopulmonary segments and then he'll find some of abnormalities .

*So, these variations of the bronchial tree may cause unexpected difficulties during bronchoscopies.

Ectopic lung lobes:

of lobes is more than normal.

*As we notice from the pic, in theRt lung there's an accessory lobe with the 3 normal lobes (so the total is 4)



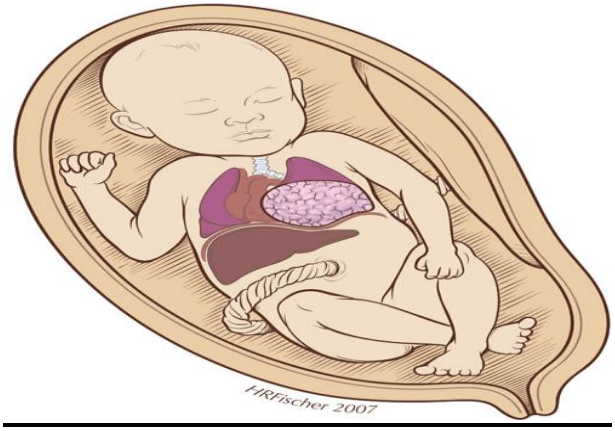
*Also ectopic lung lobes mean that some of the lung lobes are NOT in their normal position.

*It's believed that this type of anomaly does NOT affect the respiration.

((it does NOT affect the CVS *blood vessels and the circulation remain as normal*))

-**NOTE**: cardiac notch doesn't have an importance/ value clinically; it's just a deviation due to the enlargement of the heart.

Congenital cysts of the lung :



*These cysts could be either single or multiple

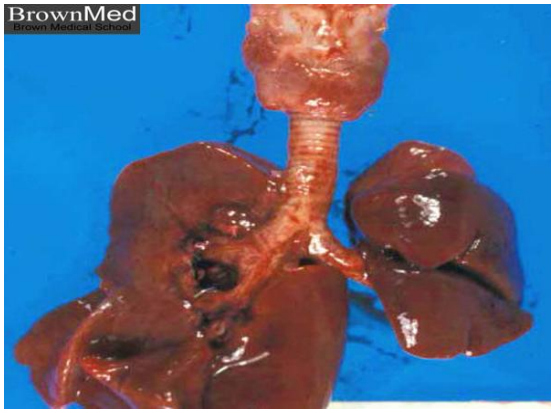
*Are formed by dilation of terminal bronchi.

*Could be multiple and appear by using the radiograph.

*Cystic structures of the lung usually drain poorly and frequently cause chronic infections (one of the complications that result from the cysts)

*Its diagnosis is easy (by using the radiograph)

Lung Hypoplasia



*In the pic, when we compare the Rt lung with the Lt one, we'll notice that the Lt lung is smaller in size and this is known as Hypoplasia of the lung.

*Very common with the diaphragmatic hernia, so in infants with **congenital diaphragmatic hernia** (CDH) the lung is unable to develop normally (there's sth prevent the lung during its development so hypoplasia will occur) and this (CDH) is more common in the Lt Side because during development there's a triangle which is wider in the Lt Side than that of the Rt side (This triangle is the cause of hernia)

**The lung is compressed by the abnormally positioned abdominal viscera.

*Characterized by reduced lung volume and most infants with CDH die of pulmonary insufficiency as their lungs cannot be expanded and are too hypoplastic to support life.

Oligohydroamnios

*Is one of the causes of Lung Hypoplasia.

*Characterized by reduced amniotic fluid which normally comes from amniotic sac and then enters GI and respiratory tract, and when it reaches the lung it'll help in the development of the lungs and alveoli. So if it's reduced, it'll lead to pulmonary hypoplasia.

*So beside Amniotic fluid's importance in the nutrition, it plays an important role in the lung development!

Lungs of the newborn infants

*As we know, babies in the intrauterine take their O_2 from the blood of their mothers. Once the mother delivers the baby, the physician will take him from his legs, invert him and hit either his legs or back leading to stimulation to the respiratory center which in turn via the phrenic nerve will stimulate the diaphragm that will get down and lungs will undergo expansion .

*Crying of the baby is the indication of the beginning of the inflation of lungs.

*Because the baby is surrounded by amniotic fluid, his skin will be slippy (bemal6es according to Dr.AlMuhtaseb :p) and this was one of the complications that physicians suffered from in the past.

*In the past, women get used to deliver in their beds because of the shortage of facilities.

*One of the problems, that the Dr faced it, was that he noticed one of the physicians after inverting the delivered baby, the baby slipped from his hands and fallen on the ground then the physician shouted that this baby was born dead!

*But if we take part of the lung and put it in water:

-If it floats, then the baby was born and breaths and cries then die

-If it sinks in the bottom of water, then the baby was really born dead

*So, this is the used way in the Forensic Medicine to differentiate between these cases!

*As a result, and according to Dr's advise, we *as physicians* should NOT lie, and should be always honest even if we make mistakes (because everybody dose mistakes!)

*And here, the problem is in the amniotic fluid, so we can solve it, as in these days, by entering the woman in a special delivery room, and the bed will be prepared in case the baby slipped, there'll be sth to support him .

GOOD LUCK

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