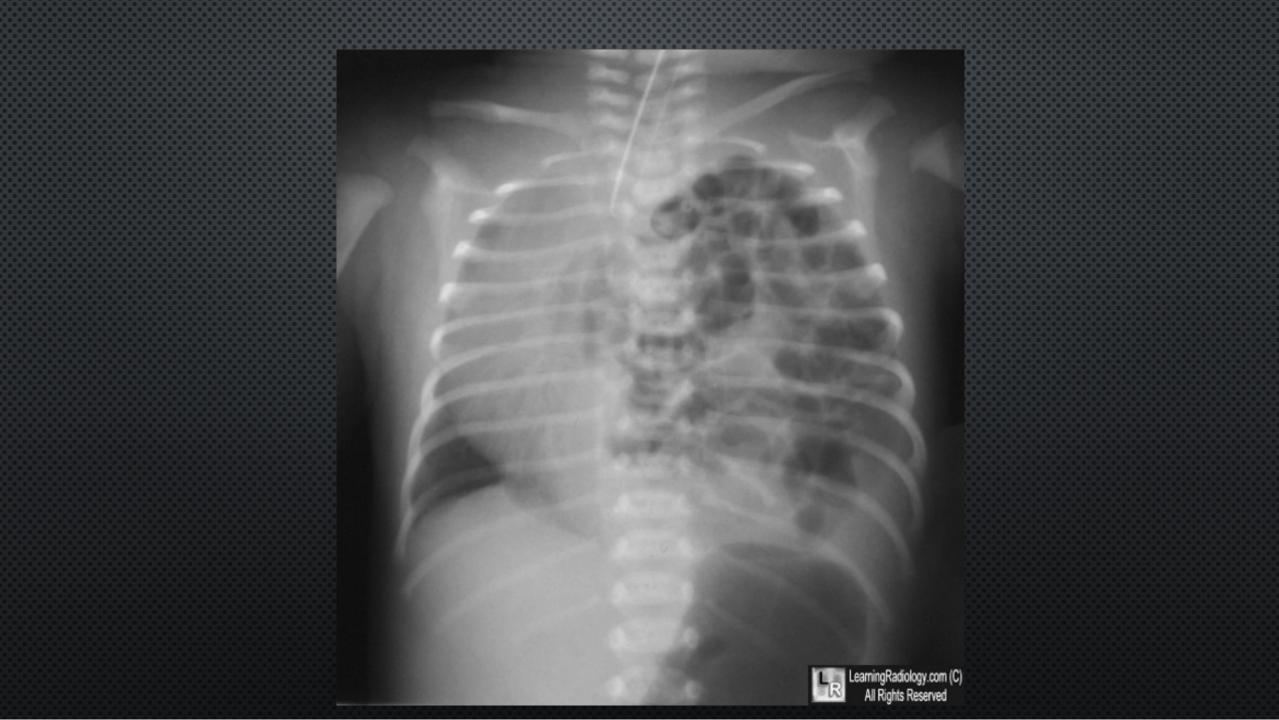
FUNDEMENTAL CASES IN RES PEDIATRIC RADIOLOGY

SYLLABUS

- CCAMA (CONGENITAL CYSTIC ADENOMATOID MALFORMATION)
- RDS (RESPIRATORY DISTRESS SYNDROM)
- CDH (CONGENIAL DIAPHRAGMATIC HERNIA)
- ROUND PNEMONIA
- ESOPHAGAL ATRASIA
- TRACHO ESOPHAGAL FISTULA
- CLE (CONGENITAL LOBAR EMPHYSEMA)







AFTER GIVING CONTRAST MEDIA ORALLY



CONGENITAL DIAPHRAGMATIC HERNIAS

There are 3 main types of congenital diaphragmatic hernia:

- posterolateral: <u>Bochdalek hernia</u>
- anterior: Morgagni hernia
- <u>hiatus hernia</u>
- Left-sided Bochdalek hernias are most common, accounting for approximately 85% of cases. Left-sided hernias, lacking the 'protective' effect of the liver may have herniation of both the small and large bowel and intraabdominal solid organs into the thoracic cavity.
- When right-sided, only the liver and a portion of the large bowel tend to herniate. Bilateral hernias are uncommon and are usually fatal due to <u>pulmonary hypoplasia</u>.

 Most congenital diaphragmatic hernias are detected either soon after birth or on antenatal ultrasound. Mortality is predominantly due to the development of <u>pulmonary hypoplasia</u>, which is thought to be due to mass effect on the developing lung. Such neonates are hypoxic and have persistent fetal circulation due to pulmonary hypoplasia and pulmonary hyper

ANOTHER CASE



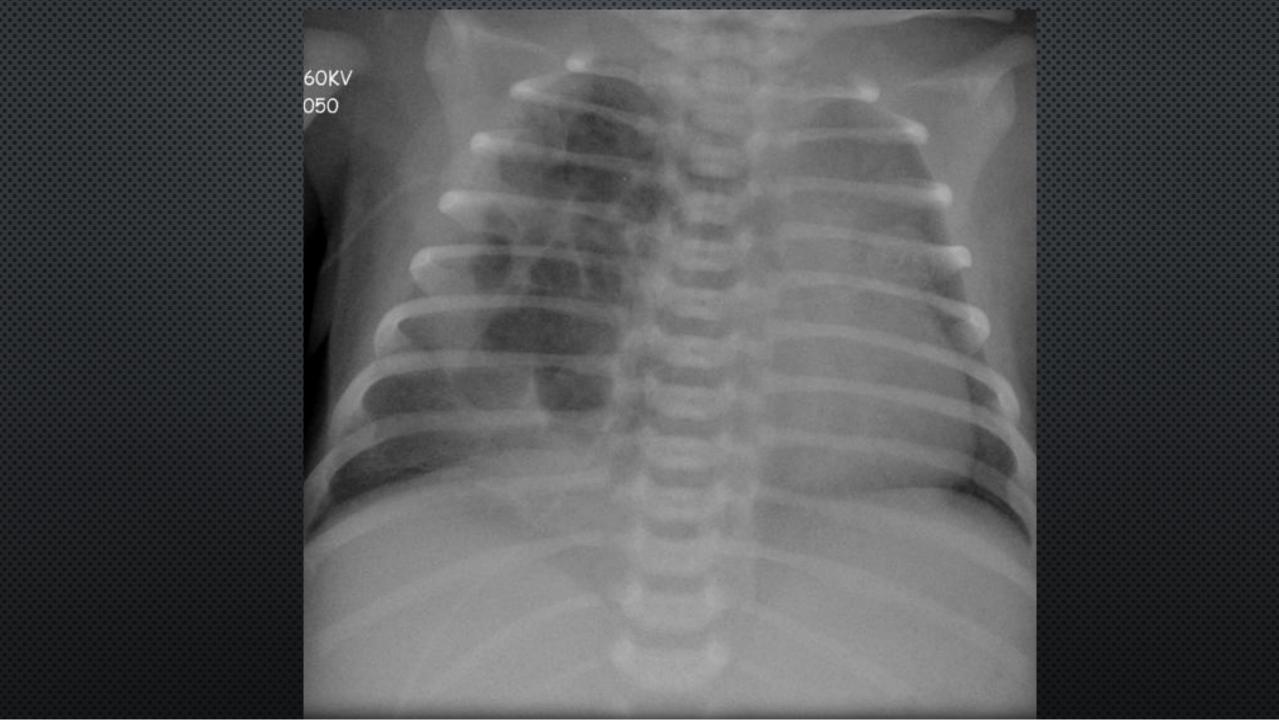
• A **Bochdalek hernia** is a form of congenital <u>diaphragmatic hernia</u>. They occur posteriorly and are due to a defect in the posterior attachment of the <u>diaphragm</u> when there is a failure of pleuroperitoneal membrane closure in utero. <u>Retroperitoneal</u> structures may prolapse through the defect, e.g. retroperitoneal fat or left <u>kidney</u>.

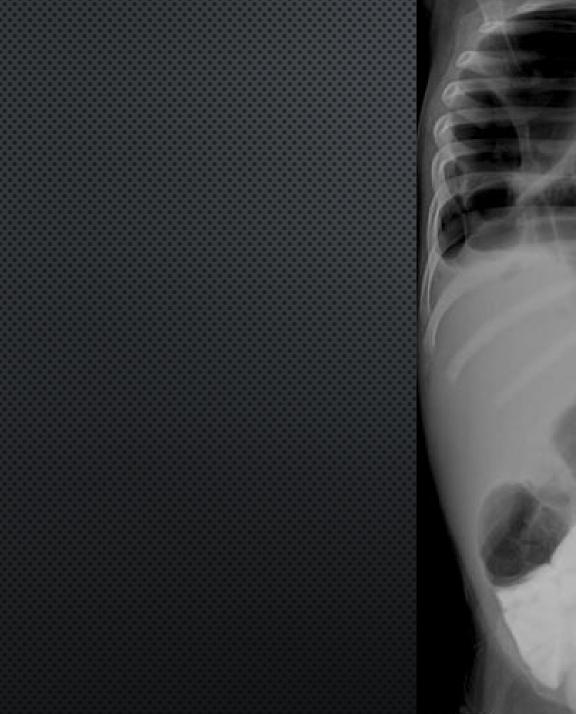
Clinical presentation

 Large hernias typically present in infancy, are usually are said to be most frequently left-sided, presumably owing to the protective effects of the liver, Complications are usually due to <u>pulmonary hypoplasia</u>

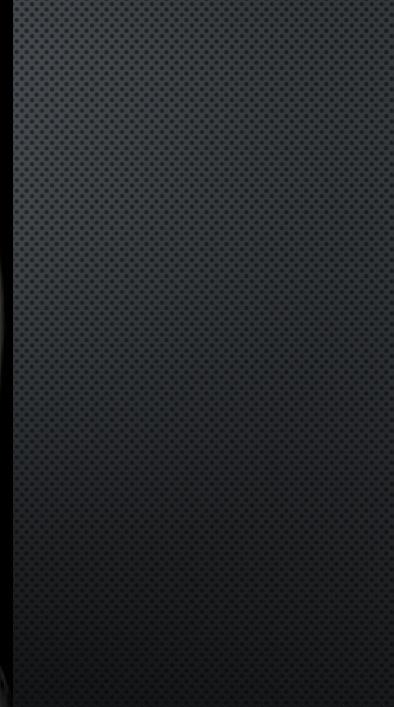
- - On conventional radiographs, the hernia may appear as a lung base softtissue opacity lesion seen posteriorly on lateral images.
 - Differential diagnosis
 - Bochdalek herniae may mimic <u>diaphragmatic rupture</u> from trauma, but you would expect to find other supportive signs of chest and/or abdominal trauma











CONGENITAL PULMONARY AIRWAY MALFORMATIONS (CPAM) CONGENITAL CYSTIC ADENOMATOID MALFORMATIONS (CCAM).

- are multicystic masses of segmental lung tissue with abnormal bronchial proliferation. CPAMs are considered part of the spectrum of <u>bronchopulmonary</u> <u>foregut malformations</u>.
- Clinical presentation
- The diagnosis is usually either made on antenatal ultrasound, or in the neonatal period on the investigation of progressive respiratory distress If large, they may cause <u>pulmonary hypoplasia</u>, with resultant poor prognosis.
- In cases where the abnormality is small, the diagnosis may not be made for many years or even until adulthood. When it does become apparent, it is usually as a result of recurrent chest infection.

FIVE SUBTYPES ARE CURRENTLY CLASSIFIED, MAINLY ACCORDING TO CYST SIZE

type I

- most common: 70% of cases ³
- large cysts
- one or more dominant cysts: 2-10 cm in size
- may be surrounded by smaller cysts
- type II

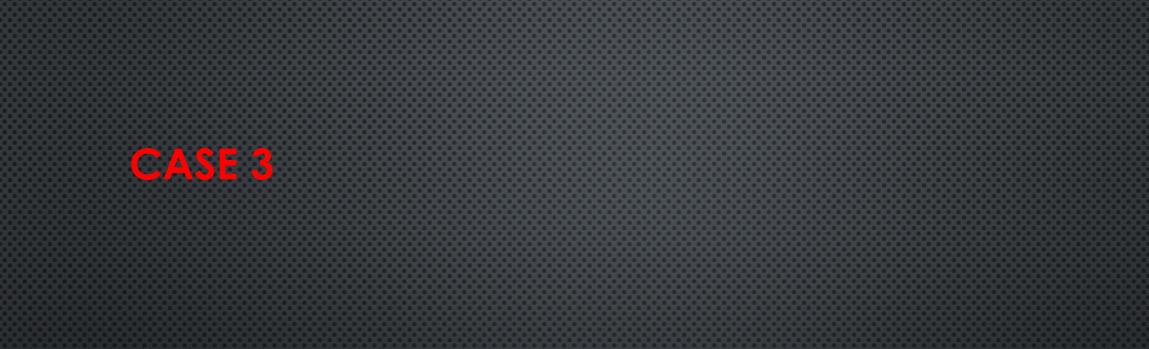
cysts are <2 cm in diameter

- associated with other abnormalities
 - <u>renal agenesis</u> or dysgenesis
 - pulmonary sequestration
 - <u>congenital cardiac anomalies</u>

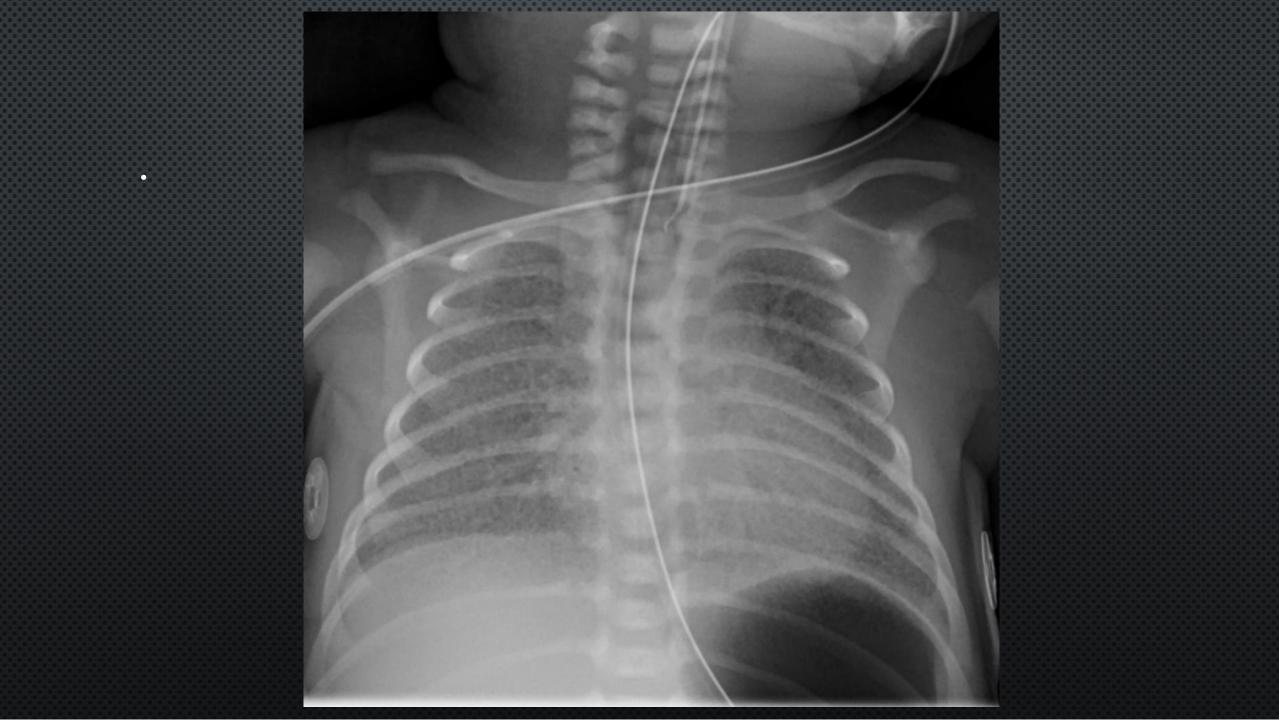
type III

- ~10% of cases
- microcysts: <5 mm in diameter
- typically involves an entire lobe
- has a poorer prognosis
- type IV
 - unlined cyst
 - typically affects a single lobe
 - indistinguishable from type I on imaging¹

- - Chest radiographs in type I and II CPAMs may demonstrate a multicystic (airfilled) lesion. Large lesions may cause a mass effect with resultant mediastinal shift, depression, and even inversion of the diaphragm. In the early neonatal period, the cysts may be completely or partially fluid-filled, in which case the lesion may appear solid or with air-fluid levels. Lesions may change in size on interval imaging. Type III lesions appear solid.



35-week gestation neonate. No complications at birth. Oxygen requirement and increasing respiratory distress requiring mechanical ventilation



RDS

- **Respiratory distress syndrome (RDS)** is a relatively common condition resulting from insufficient production of surfactant that occurs in preterm neonates.
- On imaging, the condition generally presents as bilateral and relatively symmetric diffuse ground glass lungs with low volumes and a bell-shaped thorax.

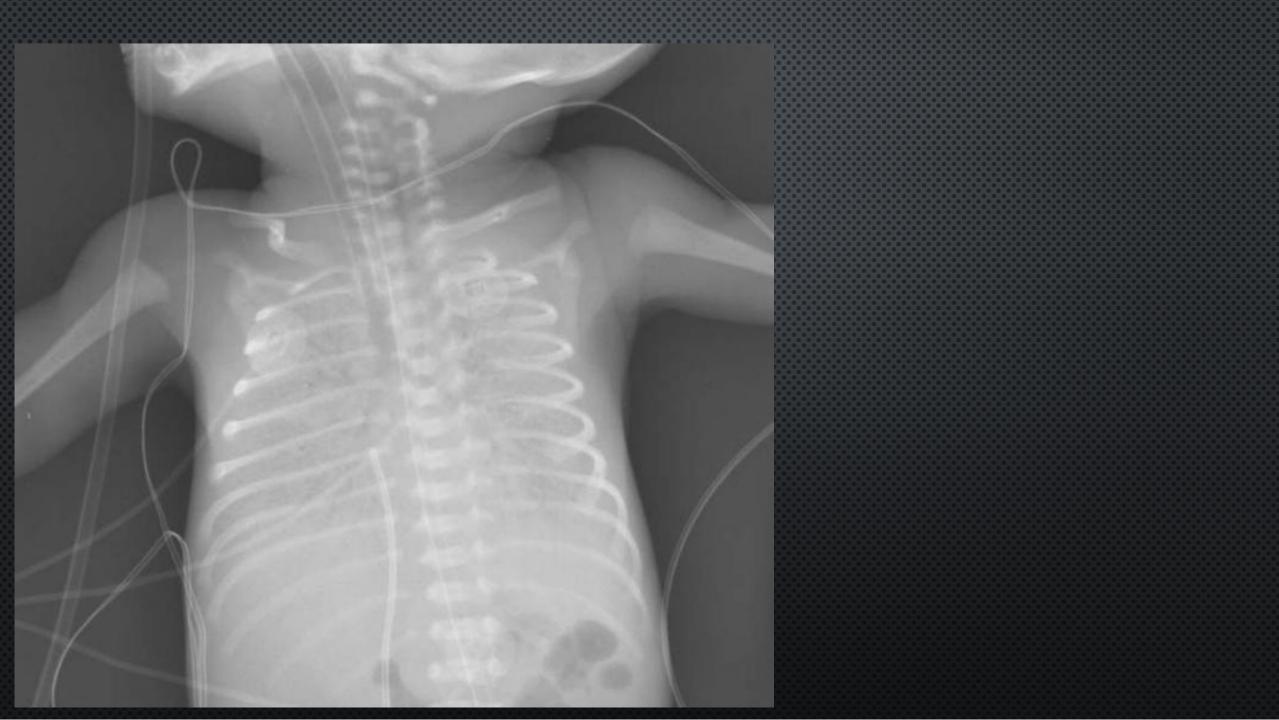
• Terminology

RDS is also known as hyaline membrane disease (not favored as reflects non-specific histological findings), neonatal respiratory distress syndrome, lung disease of prematurity (both non-specific terms), or as some authors prefer surfactant-deficiency disorder

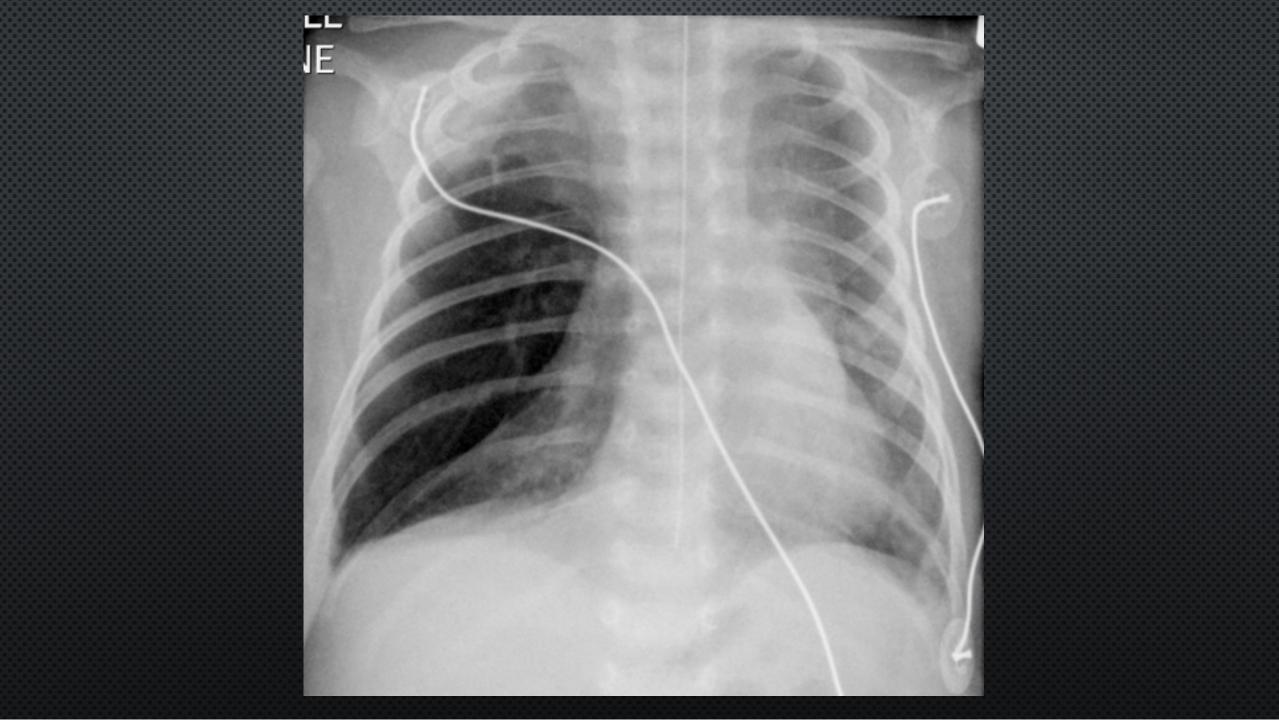
- Clinical presentation
- Respiratory distress presents in the first few hours of life in a premature baby. Signs include tachypnea, expiratory grunting, and nasal flaring. The infant may or may not be cyanosed. Substernal and intercostal retractions may be evident.

• Plain radiograph

- typically gives diffuse ground glass lungs with low volumes and a bell-shaped thorax
- often bilateral and symmetrical
- air bronchograms may be evident
- hyperinflation (in a non-ventilated patient) excludes the diagnosis
- may show hyperinflation if the patient is intubated
- RDS can be safely excluded if the neonate has a normal chest radiograph at six hours after birth.
- If treated with surfactant therapy there may be asymmetric improvement.







CONGENITAL LOBAR OVERINFLATION CONGENITAL LOBAR EMPHYSEMA)

- that results in progressive overinflation of one or more lobes of a neonate's lung.
- On imaging, it classically presents on chest radiographs as a hyperlucent lung segment with overinflation and contralateral mediastinal shift.
- Clinical presentation
- Patients typically present with respiratory distress, most commonly in the neonatal period, and usually within the first six months of life

- - Interestingly, there is a pronounced predilection for certain lobes:
 - left upper lobe: most common, 40-45%
 - right middle lobe: 30%
 - right upper lobe: 20%
 - involving more than a single lobe: 5%
 - much rarer in the lower lobes
 - Therefore, although the left upper lobe is most commonly affected, the right hemithorax is more commonly affected than the left ⁶.

- - Plain radiograph
 - Immediate postpartum period (NOT IMPORTANT FOR YOU AS 4TH MEDICAL STUDENT, JUST TO BE MENTIONED)
 - The affected lobe tends to appear opaque and homogeneous because of fetal lung fluid or it may show a diffuse reticular pattern that represents distended lymphatic channels filled with fetal lung fluid.
 - Later findings
 - appears as an area of hyperlucency in the lung with oligaemia (i.e. paucity of vessels)
 - mass effect with mediastinal shift and hemidiaphragmatic depression







TREATMENT

ROUND PNEUMONIA

is a type of <u>pneumonia</u> usually only seen in pediatric patients. They are well defined, rounded opacities that represent regions of infected consolidation.

• Epidemiology

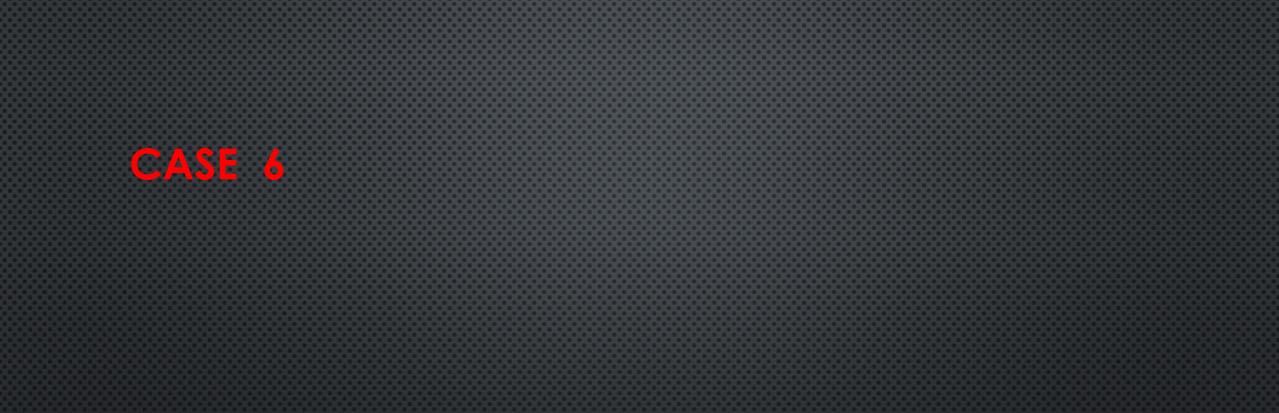
The mean age of patients with round pneumonia is 5 years and 90% of patients who present with
round pneumonia are younger than twelve Round pneumonia is uncommon after the age of
eight because collateral airways tend to be well developed by this age 3

Clinical presentation

- Patients present with symptoms of chest infection including fever, sweats, and cough. History of infective symptoms is really helpful when excluding other imaging differentials.
- The infective agent in round pneumonia is bacterial. There is no specific bacterium that causes
 round pneumonia, but since Streptococcus pneumoniae is the most common cause of chest
 infection, it is little surprise that it is the leading cause of round pneumonia⁴.

Plain radiograph

- Round pneumonias are round-ish and while they are well-circumscribed parenchymal opacities, they tend to have irregular margins. They most commonly occur in superior segments of lower lobes and in the majority of cases (98%), they are solitary ^{5.}
- <u>Air-bronchograms</u> are often present, and helpful in clinching the diagnosis.



• Newborn with direct regurgitation of milk post feed



ESOPHAGEAL ATRESIA

 Esophagus atresia is an anomaly which arises in the fourth week of the embryogenesis, at a stadium in which the trachea and esophagus should separate from each other. In case of failure of complete separation esopaghus atresia can occur.

 Clinically the neonate cannot swallow saliva, may blow bubbles and will aspirate on feeding.
 When a feeding tube is inserted it cannot be passed distally.
 A radiograph with a curled up feeding tube will confirm the diagnosis.
 Contrast swallow studies should not be performed as they can cause severe aspiration.

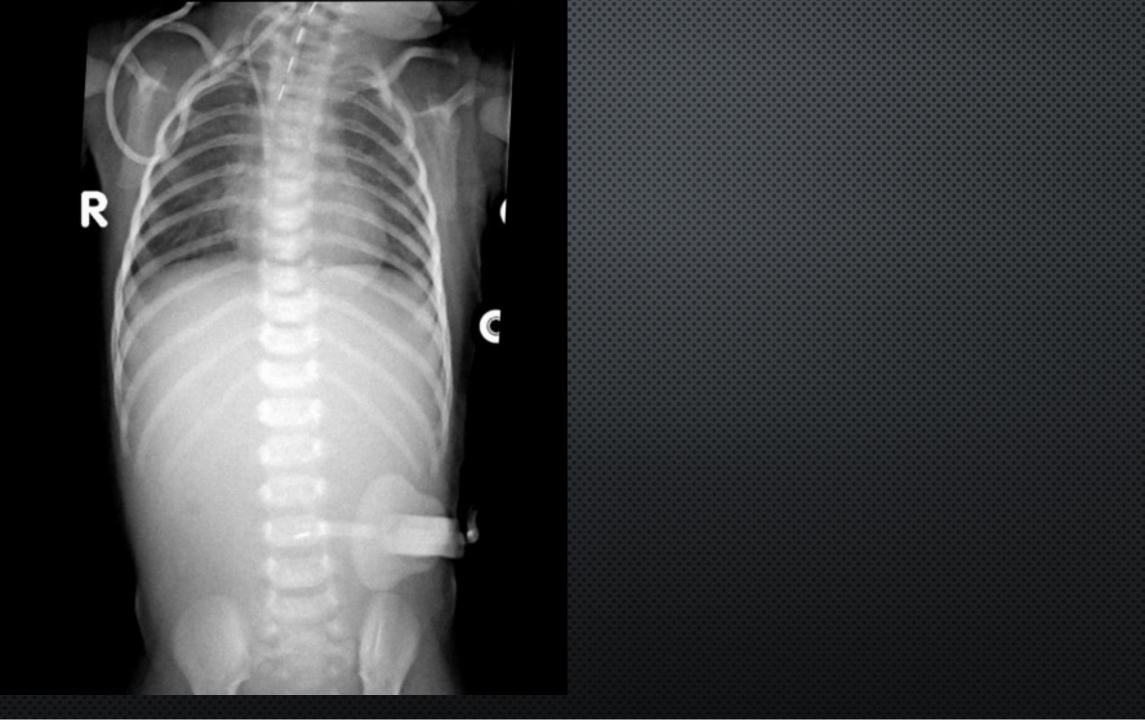
Distal tracheoesophageal fistula

No fistula

Proximal tracheoesophageal fistula

Proximal and distal fistula

- - 80% of cases a distal tracheo-esophageal fistula is present.
 - Less common is:
 - no fistula no air in stomach
 - proximal fistula no air in stomach
 - two fistulas air in stomach



- - Cases without a distal fistula can be suspected antenatally when there is a polyhydramnion and an empty stomach.
 - Always screen the radiograph for other anomalies as esophagus atresia can be part of the VACTERL association (vertebral anomalies, anal atresia, cardiovascular malformations, tracheo-oesophageal fistula, renal and limb anomalies).

