Congenital diseases of the chest

A. Tracheo-esophageal fistula. (TEF)

It is the most common congenital anomalies of the trachea, occurring in 2.4 per 10000 live birth babies.

Gross "anatomical" classification:

1. **Type A**: esophageal atresia without TEF = **8%**.
2. **Type B**: esophageal atresia with proximal TEF = **1%**.
3. **Type C**: esophageal atresia with distal TEF = **87%.**
4. **Type D**: esophageal atresia with both proximal and distal TEF = **1%.**
5. **Type** E: TEF without esophageal atresia also called H type = **3%.**

Clinical features:

They differ with different types of the anomalies, and in the most common type which is type C (esophageal atresia with distal TEF), usually and immediately after birth the neonate's mother give history of chocking, coughing and even asphyxiation and cyanosis after each feeding and there is excessive frothy salivation during time of sleeping. With less severe type the child may present late in infancy complaining of unexplained repeated chest infection with failure to thrive, malnutrition and dehydration.

In 35% it is associated with congenital cardiovascular defect and in 20% it is a part of congenital **VACTERL** syndrome.

Diagnosis:

1. **Clinical features:** feeding difficulties, excessive salivation, intermittent cyanosis and repeated or non-resolving chest infection.
2. **Radiology:** chest and abdomen X-Rays show large amount of air in the esophagus, stomach, and small bowel, (**in type C**).
3. **Failure of normal advance of N/G tube in the esophagus** under fluoroscopic monitoring.
4. **Contrast esophagography** show full anatomical demarcation of the defect, in type C the esophagus appears as proximal pouch.

Treatment:

1. Put the neonate in the **incubator** to avoid hypothermia.
2. **Semi recumbent position** (elevation of the head and chest 30°).
3. **Oxygen supplementation** for those patients with hypoxia or cyanosis due to aspiration or repeated chest infection.
4. **Frequent suction** of saliva and secretion from the mouth and oropharynx to minimize aspiration.
5. **Surgical repair**. (in type C by RT extrapleural thoracotomy, with fistula is divided and over sewn, the proximal and distal part of the esophagus are approximated and anastomosed together. In type H if high type (cervical type) it approached by RT cervical incision and dissection done until reach the fistula which divided and over sewn, while in low type (thoracic type) can be managed by thoracoscopic ligation.

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B. Pulmonary sequestration.

(sequestration means isolation or separation)

It is a pulmonary anomaly in which the abnormal part of the lung has systemic arterial supply and abnormal bronchial connection, they are invariably located in the base of the lung with 3:1 male to female ratio. It is a congenital anomaly in which a portion of the developing lung is separated completely from the remaining lung parenchyma and tracheo-bronchial tree, with its blood supply is derived from anomalous systemic vessel rather than pulmonary circulation.

Types:

1.Intralobar (75%): when the lung defect located within the visceral pleura of the lobe, the systemic arterial supply come from thoracic aorta 74%, abdominal aorta 19%, intercostal arteries 3% and multiple source in 20%. The venous drainage is usually to the pulmonary veins and may be to the systemic veins, the entry of the vessels is away from the hilum, it mostly found in the posterior segment of LT lower lobe and it became symptomatic at adolescence or young adult as repeated bouts of chest infection and hemoptysis, rarely associated with other congenital anomalies.

2.Extralobar (25%): located outside the visceral pleura of the lobe, and has its own separate visceral pleura, 90% are rounded, smooth soft mass, above the dome of the LT hemidiaphragm, and sometimes intrapericardial or intradiaphragmatic or subdiaphragmatic retroperitoneal, it may associate with other congenital anomalies especially diaphragmatic hernia, pericardial cyst, esophageal atresia and cardiac defect. the venous drainage mostly to the systemic circulation Azygos and Hemiazygos veins and in 20% to the pulmonary circulation, it has some tendency for malignant neoplasm changes.

Diagnosis:

1. **Tumor markers:** marked elevation of the Serum CEA and Serum CA19-9.
2. **Chest C-T Scan with i.v. contrast and C-T Scan angiography of the chest and abdomen:** to delineate the blood vessels (the anomalous systemic arterial blood supply and anomalous systemic or pulmonary venous drainage).
3. **Chest MRI:** better demarcation of the tissue planes and better differentiation of the normal lung tissue from sequestrated lung tissue.
4. **Barium esophagogram** to detect any esophageal communications to the tracheo-bronchial tree i.e. TEF (tracheo-esophageal fistula).

Treatment:

If the sequestrated part of the lung is huge, cystic and cause hemodynamic compromise and respiratory embarrassment of the infant, so needs **rapid decompression by needle aspiration or chest tube**, prior to induction of general anesthesia for the proposed surgery.

Surgery is done by postero-lateral thoracotomy with the **removal of the cyst (sequestrated part) itself** or by the removal of the segment or the lobe that contained the sequestrated lung parenchyma, the most common surgical procedure is lobectomy.

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C. congenital lobar emphysema.

Congenital lobar emphysema is not uncommon cause of respiratory distress in the newborn and is characterized by overexpansion of a pulmonary lobe due to alveolar overdistension.

Etiology and pathogenesis:

It is not clear in the majority of the cases, however both bronchial cartilaginous dysplasia and polyalveolar lobes are the pathologies present in 25% of the cases for each pathology. extrinsic compression of a lobar bronchi can occur from an intrathoracic mass such as a vascular or cardiac anomaly, teratoma, vascular ring or patent ductus arteriosus. Even the end stage of respiratory distress syndrome in a neonate with a chronic pulmonary interstitial emphysema can evolved into congenital lobar emphysema.

A variant of bronchomalacia can cause congenital lobar emphysema with focal cartilaginous deficiency of the tracheobronchial tree resulting in regional airway collapse with expansion and air trapping leading to emphysematous changes.

Twenty percent of the associated congenital malformations are related to cardiovascular system include cardiac anomalies, patent ductus arteriosclerosis, ventricular septal defect and tetralogy of Fallot. moderate distress present initially until the lobe gradually enlarges causing cyanosis by continued compression of the uninvolved lobes and lung due to continued expansion of the affected lobe. Severe life-threatening distress is rare but may require immediate surgical intervention. Bronchoscopy and positive pressure ventilation can exacerbate the underlying pathophysiology of congenital lobar emphysema creating a life-threatening situation which can only be relieved by rapid thoracotomy to decompress the hyperinflated lobe.

Diagnosis:

1. **Clinical features:** over one half are diagnosed by one month of age and the presentation after six months is unusual, the neonate presented with features of respiratory distress, mild to moderate dyspnea and cyanosis is not so frequent, the ipsilateral hemithorax appear over expanded by the emphysematous lobe with restricted movement on respiration, there is diminished air entry on the affected side with hyperresonance on percussion looks like features of pneumothorax, the trachea and mediastinum appear shifted to the contralateral side.
2. **Chest X-Ray:** is sufficient to diagnose a large emphysematous lobe with collapse atelectasis in the ipsilateral lobe or lobes. the upper lobes are most commonly involved with the following incidence, left upper lobe 42%, middle lobe 35%, right upper lobe 21% and bilateral 20%.
3. **Chest C-T Scan:** when still there is doubt about the accurate diagnosis based on clinical examination and chest X-Ray, the affected lobe appears more translucent due to decreased densities of the broncho-vascular markings with loss of the normal lung architecture. The emphysematous lobe may appear herniated across the mediastinum to the contralateral hemithorax, it is usually causes collapse atelectasis of the adjacent lobe and in more severe cases it may even causes a compression of the contralateral lung.

Treatment:

1. The neonate is **managed in the neonatal intensive care unit** (critically ill patients), put in the **incubator** with **semi recumbent position** so that the head and the chest are elevated to 20-30˚.
2. **Oxygen supplementation** may be given to cyanosed patients or those with severe respiratory distress.
3. **Associated congenital malformations** is dealt with accordingly.
4. Surgery is the definitive treatment done under general anesthesia by thoracotomy and lobectomy of the involved lobe after optimization of the general and respiratory conditions of the patient.

Early diagnosis requires high index of suspicion and familiarity with the representation since they are difficult to detected, whereas early diagnosis is amenable to an uncomplicated surgical resection, a delay in the diagnosis can be associated with substantial morbidity.