Congenital diseases of the chest

D. Bochdalek hernia.

It is a congenital malformation of the diaphragm in which there is a variable size of posterolateral diaphragmatic defect and herniation of abdominal contents through this defect. The diaphragmatic defect is caused by failure of pleuro- peritoneal canal fusion during the fetal development. More than 80% occurs in the left hemithorax and typically through a small circular deflect, only 10% have a true hernial sac. Infants with Bochdalek hernia diagnosed at birth, accounting 1/4000 of live birth and have relatively poor prognosis because those patients present early in life have more severe malformations.

Etiology:

During the development of the embryo in the intra-uterine life and at the 10^{th} week of gestation the abdominal organs may herniate into the thorax through a persistent defect in the postero-lateral part of the hemidiaphragm, the defect is the result of failure of complete closure of pleuro-peritoneal canal. The pressure exerted by the abdominal viscera in the thoracic cavity results in impaired growth of the ipsilateral lung.

This type of diaphragmatic hernia is commonly associated with additional anomalies including congenital heart defects, hydronephrosis, renal agenesis, intestinal atresia, extralobar sequestration and other anomalies.

The other elements that contributes to the pathogenesis of this anomaly is pulmonary hypoplasia and hence pulmonary hypertension, the timing of the presentation is depend on the degree of pulmonary hypoplasia which is the main cause of morbidity and mortality associated with this disorder that results from severe hypoxia and acidosis.

Clinical features:

It may result in a wide spectrum of respiratory symptoms that varied from life-threatening respiratory compromise as early as the first few hours or days of life, to the degree that occasionally the defect may result in respiratory distress or feeding intolerance later in infancy or childhood or may be the defect is identified on radiograph obtained for unrelated reasons.

Diagnoses:

1. **Chest X-ray:** characteristically demonstrates abdominal contents in the chest with mediastinal deviation away from the affected side, the visualization of nasogastric or orogastric tube above the diaphragm also support the diagnosis.
2. **Contrast study:** including barium swallow, meal, follow through and enema which demonstrate the presence of different part of the bowel in the posterior-lateral aspect of the chest.

3. **Chest CT scan with or without oral contras:** that's barium swallow, meal and enema this is done with when other investigative tools failed to reach the diagnosis.

Treatment:

1. The patient should be admitted to the neonatal intensive care unit and managed in the incubator in semi recumbent position. (head and chest elevated 20-30˚).

2. Oxygen supplementation may be given to patients with severe respiratory distress or cyanosis.

3. Stop oral feeding and insert nasogastric or orogastric tube for bowel decompression.

4. Surgery there are two types of surgical approaches to repair this type of hernia, the abdominal approach by upper paramedian or subcostal incisions, this is indicated for almost all patients and the thoracic approach which is done by postero-lateral thoracotomy and is indicated for two conditions, the right sided Bochdalek hernia or late presentation during the period of young childhood.

E. Congenital chest wall deformities

Children with congenital chest wall anomalies may present from infancy to adolescence with cosmetic concerns, psychological disturbances and physical disabilities. The most common anomaly of the chest wall includes pectus excavatum and pectus carinatum, with many other rare congenital anomalies.

A. pectus excavatum

Introduction:

Abnormal growth, lengthening, rotation or increase elasticity of the costal cartilages lead to relatively common depression of the sternum known as pectus excavatum, the clinical findings usually begin to manifest by 2 to 3 years of age. Two types of repair can be done an opened repair called Ravitch procedure and a less invasive repair called Nuss repair. The type of repair is chosen based on the surgeon's experience and the complexity of the deformity.

A study of biomechanical properties of the costal cartilage from children with pectus excavatum shows decreased tension, compressible, and flexure with disrupted typeⅡ collagen patterns in the deep zone of the cartilage.

The incidence of pectus excavatum is approximately **1∶400** live births and the male∶ female ratio is about **5∶1**, there is high association of this congenital anomaly with Marfan's syndrome, Ehler-Danlo's Syndrome and congenital heart disease.

Clinical Futures:

Most of the children appear active and healthy and complaining chiefly of visible anatomical defect, they may have non-specific parasternal chest pain and decreased subjective exercise tolerance, they also have frequent respiratory tract infection and exercise induced asthma. Some of the patients at the teenage may have embarrassment reactions, social anxiety, depression and suicidal thoughts. **On examination** they have visible depression of the lower sternum with right sided rotation, sloped ribs, rounded shoulders, protuberant abdomen and laterally displaced cardiac point of maximal impact.

Diagnosis:

1. **Chest X-ray:** it allows the measurement of severity index and the extent of abnormal calcification of costal cartilages.
2. **Chest CT scan:** this study provides precise information regarding cardiac displacement, lung volume and more importantly the most accurate measurement of the depth of the sternal defect and its relationship to the overall width of the thorax.
3. **MRI:** can give a better radiological assessment.

 # severity index: two methods for the measurement of this index these are

A. **Haller's index:** transverse diameter of the chest/A-P diameter of the chest.
B. **Sternal index:** A-P diameter of chest cavity/height of patientX100.

Indications for surgical repair:

1. **Anatomical**: when there is significant anatomical defect that evident on clinical and radiological examination.
2. **Physiologic**: when there is impairment of cardiac and respiratory function.
3 .**Psychological**: development of psychological disturbances.

Surgical treatment

A. Modified Ravitch repair: consist of elevation of the depressed lower sternum by subperichonderial division of the lower 4 costal cartilages bilaterally with a transverse anterior wedge osteotomy of the sternum which is then supported by stainless Steel rod.

B. Nuss procedure: through small bilateral incisions a convex steel bar was surgically placed under the sternum to allow repositioning and remodeling of the deformed costal cartilages and sternum after keeping the bar in place for two years to allow distraction and splinting of the costal cartilages as well as the rotation of the sternum to a more neutral position the bar was removed.

B. Pectus carinatum

Introduction:

It is a congenital deformity of the chest wall with anterior protrusion of the sternum as a result of rotation and lengthening of the coastal cartridges resulting in sternal protrusion. The incidence is about **0.06** of live births, males are affected more than females and the usual presentation is at a teenage, it also associated with other skeletal deformities.

Defective and elongated cartilages is responsible for pectus carinatum as well as excavatum, rapid growth of the costal cartilages with the sternum displaced forward this will cause pectus carinatum, and if these cartilages displaces the sternum posteriorly then pectus excavatum occurs.

Since no significant cardiopulmonary abnormalities can be demonstrated with this lesion of the chest, pectus carinatum remains a clinical diagnosis and no further diagnostic tests are required. CT scan of the chest may be indicated to exclude any other intrathoracic abnormality.

Clinical Futures:

Symptoms: parasternal pain, frequent injuries, exercise intolerance and decreased work capacity.
Examination: external elevation/rotation of the sternum, unilateral or bilateral costal cartilages elevation and barrel chest.

Surgical treatment:

Surgical repair includes:

1. Transverse or midline incision along the sternum.

2. Creation of skin subcutaneous and muscle flaps.

3. Subperichonderial excision of all elevated costal cartilages.

4. Subperichonderial contralateral costal cartilage resection (if unilateral overgrowth).

5. Anterior sternal osteotomy to depress sternum to neutral position.

6. Bone wedge inserted inside the anterior sternal osteotomy to hold sternum in a neutral position.

7. Wound closure with closed drainage.