Congenital Diaphragmatic Hernia

Presented By Nick Mancuso

Objectives

Overview of CDH

- Embryology
- Case Study

CDH

• What is it?

 CDH is when the contents of the abdomen rupture through the diaphragm into the thoracic cavity





Classification

- 6 major types of CDH
 - Retrosternal or retrocostosternal
 - Posterolateral defects
 - Eventration
 - Esophageal hiatus hernia
 - Extensive defects of hemi-diaphragm
 - Herniation into the pericardium

Causes of CDH

- There are three known causes of CDH
 - As discussed failure of the diaphragm to fully develop
 - Return of the midgut to the abdominal cavity from the umbilical stalk
 - Lung formation

- Normal Development
 - Cranial-caudal folding basically establishes the precursors of all the structures and cavities these structures will invest in the normal thoracic cavity



- Diaphragm formation
 - Body folding brings the septum transversum caudal to the pericardial cavity
 - Five major components of the diaphragm
 - Septum transversum
 - 2 pleuroperitoneal membranes
 - Esophageal mesentery
 - Body wall



Normal development o the diaphragm, all five tissues present

• Abnormal Diaphragm Formation

 Any failure of these tissues to develop or weakness in the connection could lead to CDH following further embryo development

- Midgut Formation
 - Midgut grows faster and larger than the embryo can sustain so it extends into the umbilical stalk
 - Around the tenth week it returns to he abdominal cavity to continue formation and folding of the GI tract

- Midgut
 - Premature return of the midgut, prior to formation of the diaphragm will result in CDH due to the increased pressure from the abdominal cavity and the contents will rupture into the thoracic cavity
 - Also, appropriate return of the midgut with delayed diaphragm formation will result in CDH

- Lung Formation
 - If the lungs do not develop appropriately later in fetal development, the mechanical pressure from the abdomen, even with a fully formed diaphragm will cause the abdominal contents to push into the thoracic cavity, increasing pressure on the thoracic viscera
 - Bronchial and alveolar development lag behind much other organ development

Why is CDH bad?

- Increased pressure combined with smaller room for development can result in pulmonary hypoplasia (defective/incomplete development), the severity depending on the time of herniation
- Also, this will result in a shifting of the other thoracic viscera which can hamper development and function

Case study

- Symptoms of CDH
 - Subcostal and/or sternal retractions
 - Scaphoid abdomen with lack of bowel sounds
 - Cyanosis
 - Respiratory distress
 - Possible polyhydramnios
 - Decreased lung sounds

- Treatment
 - Requires surgical correction, a number of different approaches are used entering from the thoracic or abdominal cavity
 - Herniated organs are pulled/pushed into abdominal cavity, and herniations are repaired

- 22 hour-old male infant born at term weighing 3,200 grams
- Uncomplicated birth
- Suffering dyspnea (respiratory distress)

- Examination
 - Infant had subcostal and sternal retractions, scaphoid abdomen with a lack of bowel sounds, and cyanosis relieved with oxygen
 - X-rays showed gas-filled intestinal loops in left thorax with mediastinal shift to right with left lung hypoplasia

Diagnosis CDH through foramen of Bochdalek

Treatment

Infant underwent surgery, but left lung did not expand and patient ultimately passed away

Statistics

- CDH comprises ~8% of all major fatal congenital anomalies, and is responsible for 1 in 50 neonatal deaths in first three days of life
- 1 in every 2,000 to 4,000 live births

Questions?