CONGENITAL DIAPHRAGMATIC HERNIA ROUND TABLE
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CDH: Anatomy
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Congenital Diaphragmatic Hernia (CDH)
- A birth defect where a hole in the diaphragm permits some or all of the abdominal organs to migrate into the chest cavity

Incidence
- CDH is present in approximately 1 in 2500 births annually
  - Increased likelihood of occurring with subsequent pregnancies of 1 in 50
  - Increased likelihood of a comorbid heart defect (1 in 5) and with genetic abnormalities (1 in 10)

Historical Overview - From 1850s to today
CDH: Anatomy

**Bochdaleck**
- Posterolateral diaphragmatic hernia
- 99% are on the left side

**Morgagni Hernia**
- Retrosternal or parasternal hernia
- 2% of all CDH cases
- Abdominal contents herniate via the foramina of Morgagni (adjacent and posterior to the xiphoid process)

★ **Diaphragm Eventration:**
- The diaphragm is thin but intact and can allow abdominal contents to protrude into the chest cavity

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**Pulmonary Sequelae - The Real Problem**

- **Hypoplasia**
  - Abdominal contents in the chest cavity during fetal development cause underdevelopment of the lungs causing respiratory distress or worse

- **Hypertension**
  - Blood vessels in the lungs do not form correctly and the lung pressures after birth fail to decrease physiologically
  - Fewer blood vessels form due to pressure on the lungs by abdominal contents pushing on them during development

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**REFERENCES**

Al-Salem, AH. An Illustrated guide to pediatric surgery. 2014. 329-344.


CDH: Pre Operative Management
Linda Zekas, APN, RN, CPNP, CNNP, WOCN

CDH: Prenatal Diagnosis
- Usually noted on 20 week US (A/B)
  - Stomach seen on same view as heart (A)
  - Done sequentially throughout remainder of pregnancy
- +/- MRI imaging (C)
  - Other anomalies
  - Prognostic tools
    - Fetal lung volumes
    - Lunghead ratio
- Historically, key things to note:
  - Right vs. left side
  - Liver up or down
- Limitations of prenatal imaging
- Additional prenatal testing: ECHO

CDH: Delivery Room Management
- May present with mild or profound respiratory compromise
- Limit bag/mask ventilation
- Intubate to prevent gaseous abdominal distention
- Place gastric tube ("life line")
- Pre/postductal CO2 saturation
- Physical exam findings:
  - Scaphoid abdomen
  - Barrel chest
  - PMI shifted
  - Unequal breath sounds
### CDH: Postnatal Presentation / Diagnosis

- Respiratory deterioration over the course of hours to days in the neonatal period
- Respiratory distress and feeding difficulties that develop over the course of weeks to months
  - Respiratory and abdominal symptomology (i.e., feeding intolerance) may be fluid
  - Think of CDH as door allowing abdominal contents to migrate in and out
  - Ease with which this can happen dependent on size of defect
  - May develop intestinal obstruction
- Incidental finding on radiologic imaging +/- clinical symptomology
- In general, later presentation/diagnosis imparts better short/long-term outcomes

### CDH: Preoperative Management

Historically, repair of the defect was considered the primary objective upon presentation. Current thought is to optimize pulmonary hypertension management which is now seen as key to decreasing some of the morbidity and mortality associated with CDH repair.

- **Pulmonary hypertension management**
  - Limit stimulation
  - +/- ECHO while in pulmonary hypertensive crisis
  - Medications available: iNO, milrinone, sildenafil, prostacyclines, bosentan, treprostinil
  - +/- serial BNP's to follow trend
  - ECMO for most severe cases of pulmonary hypertension

- **Preoperative testing/preparation**
  - Babygram or CXR/KUB (older children)
  - ECHO
  - Labwork (Chem 8, CBC, coagulation profile)
  - 1 unit PRBC's on hold to the OR
  - Remember, gastric decompression is key to preventing sudden respiratory/cardiovascular compromise

### CDH: Intraoperative Management

Lisa Iamiceli, CPNP, MSN, RN, CNS
CDH: Intraoperative Management

- Minimally Invasive Technique vs. Open Repair
  - Classic approach: laparotomy via subcostal incision
  - Reduction of visceral contents into the abdomen
  - Primary vs. patch closure
  - Timing of repair
  - Advantages and disadvantages to minimally invasive approach

CDH: Intraoperative Management

- Historical Repair of CDH
  - Previously done immediately after birth
  - Anticipated improvement in ventilation with reduction of visceral organs from the thoracic cavity
  - No improvement in rate of survival with early repair
  - 1980's: more common to delay repair
  - Current practice: improvement of lung physiology and ventilator status prior to repair
CDH: Intraoperative Management

- APN role in the operating room
- Continuity of care
- Communication with the family, prenatal consult to postnatal care
- Surgery to NICU handoff
  - Post op recommendations

References


CDH: Postoperative Management

Jennifer Reitsma, MSN, APRN-CPNP
CDH: Postoperative Care

Postoperative report:
- ECMO
- Estimated blood loss
- Type of repair
- Degree of pulmonary hypoplasia
- Respiratory support (prior to OR case vs postoperative requirements)
- Chest tube
- Thoracotomy vs thoracoscopy
- Type of hernia repair
  - Gore-tex patch vs sutured repair
- Lines/vascular access
- Dressings
- Vital signs

CDH: Components of a chest tube system

- Collection chamber
- Water seal chamber
- Dry suction control regulator
- Air leak monitor

CDH: Restore negative pressure in the pleural space

The depth of the water in the suction bottle determines the amount of negative pressure that can be transmitted to the chest, NOT the reading on the vacuum regulator.
From bottles to box

- Suction control bottle
- Water seal bottle
- Collection bottle

To suction

- Suction control chamber
- Water seal chamber
- Collection chamber

From patient

CDH: Chest Tube - From box to bedside

- Water seal is a window into the pleural space
- Not only for pressure
- If air is leaving the chest, bubbling will be seen here
- Air leak meter (1-5) provides a way to "measure" the leak and monitor over time – Is the leak getting better or worse?

CDH: Chest Tube - Monitoring Air Leak

- Water seal is a window into the pleural space
- Not only for pressure
- If air is leaving the chest, bubbling will be seen here
- Air leak meter (1-5) provides a way to "measure" the leak and monitor over time – Is the leak getting better or worse?
CDH: Chest Tube - Setting up the drain

- Add water to the 2cm level in the water seal chamber, and to the 20cm level in the suction control chamber (unless a different level is ordered)
- Connect chest drain tubing to thoracic catheter
- Connect the drain to vacuum, and slowly increase vacuum until gentle bubbling appears in the suction control chamber

CDH: Chest Tube - Typical Use

- Underwater seal (UWS) will typically be used for air that needs to evacuate pleural space.
- Suction will typically be used for fluid that needs to be evacuated from the pleural space.

CDH: Chest Tube Connections
CDH: Chest tube - Closed system

Avoid leaks; check the system if there is bubbling

- Dressing must be secure
- Connections must be tight
- Zip strip gun settings

CDH: Thoracotomy vs. Thoracoscopy

Thoracotomy
- Most painful incision possible.
- Long term complications of winged scapula, scoliosis.

Thoracoscopy
- Smaller incisions
- Less pain, shorter length of stay
- Longer surgical time
- Reliant on energy source for vessel ligation
### CDH: Postoperative Assessment

**Vital signs**
- HR
- Temperature
- Oxygen saturation
- BP
- Pain assessment (use the appropriate pain scale for age)

**I&O**
- Chest tube output
- UO

**Laboratory and studies**
- CXR postoperatively including abdomen
- LFT’s if liver involved
- Pancreas

### CDH: Discharge Teaching

1. Follow up appointments
2. What to call with...
3. Contact numbers
4. How to position the child
5. Dressing care
6. Bathing

### CDH: Postoperative Management

**REFERENCES**

CDH: Long Term Follow-up
Elin Öst, RN, PhD Student

Approximately 87% of CDH survivors have long lasting associated morbidity. Many centers have initiated standardized follow-up programs to ensure that all morbidity areas are covered. Those areas are neurodevelopmental, pulmonary, gastrointestinal and musculoskeletal outcome. Surgical complications in CDH survivors are common and can occur asymptomatic many years after the repair. Early recognition of symptoms may increase survival and prevent secondary mortality. Neurodevelopmental outcome
Children with CDH more often suffer from neurodevelopmental and neuro-functional sequelae than the healthy population, probably as a consequence of perinatal and neonatal hypoxiaemia. Gestational age has been identified as one isolated predictor for neurodevelopmental delay, where preterm, late preterm and near preterm children with CDH had an increased risk. Especially motor performance seems to be affected by gestational age.
Children with CDH score normal IQ when testing with Wechsler Intelligence Scale for Children (WISC) but seems to be at risk for attention and concentration deficits. However children with CDH treated with ECMO has shown lower IQ scores than those without ECMO treatment. The vast majority of all children with CDH attend normal school but as many as half of them are in need of extra support in school. Since children with CDH may be at risk for behavioral problems, cognitive functioning should be tested at standardized follow-up. This risk is not related to the severity of the malformation why all children with CDH should be included in the follow-up.

Sensorineural hearing loss has been described with a varied range of cases, 3-60% among children with CDH. Pulmonary outcome
Even though new strategies for therapy aim to minimize pulmonary sequelae induced by barotrauma, pulmonary morbidity is commonly described. With asthma symptoms, recurrent respiratory tract infections and an impaired pulmonary function. In long-term follow-up in adult CDH survivors nearly 50% showed an impaired pulmonary function when testing. In a study where CDH survivors viewed their personal fitness, a feeling of being less fit than peers and having a negative attitude towards exercise was described. A higher level of performance leads to a less perception of dyspnea and effort.
Gastrointestinal outcome
Gastroesophageal reflux (GER) is a common consequent of CDH. The prevalence of GER varies among different studies, 20-84% during the first year of life. In adults with CDH 54% showed endoscopic or histological GER. Gastroesophageal reflux can cause comorbidities such as failure to thrive, oral aversion, pain and worsened pulmonary morbidity. Symptoms for detection of esophagitis has been described as poor and many patients have been completely asymptomatic while other have had heartburn or regurgitation. Failure to thrive has been described as a cause of GER but can also be due to prolonged endotracheal intubation and increased caloric requirement because of pulmonary morbidity.
Musculoskeletal outcome
Chest wall deformaties, often in form of an asymmetric pectus excavatum or flat chest, are present in about 50% of all children with CDH. Scoliosis is present in 12% of all children with CDH and correlates with a large defect. In adult survivors with CDH 27% has scoliosis.
Health Related Quality of Life

There are only a few QOL studies in children with CDH, with ambiguous results. Most of them came to the conclusion that the youngest children scored lower than the general population. However, parents normally score lower QOL when answering on behalf of their children.

Parental stress

Parents of chronically ill children, especially mothers, report higher levels of anxiety and depression than parents of healthy children, whereas problems in daily life and parenting stress are significant predictors. Attention for parent well being, for example by use of screening-tool, all follow-ups for children with CDH could serve as a start of communication about the parental need.