Management of Congenital Diaphragmatic Hernia

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5/15/13
• 5 month old BB admitted for several days of non-bilious, non projectale vomiting
• PMH: None
• PSH: None
• Allergies: NKDA
• Second child, vaginal delivery, normal pregnancy, Apgar 8
• Family Hx: Inguinal hernias in mother & sibling.
• VS: T 98.3; BP 108/66, HR 144, RR 44, sat 100% on RA

• Labs:
  – CBC: 6.5<9.6/29.6>190
  – AST/ALT: 33/21; Alk phos 245; Albumin 4.1
UGI series: right-sided diaphragmatic hernia.

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CXR: Retrocardiac and right basilar opacity with lucency. Suspicious for diaphragmatic hernia. The left lung is clear.
Postop day #1
Postop day #1 (2nd procedure)
Postop day #3 (2nd procedure)
Postop day #1 (third procedure)
Congenital Diaphragmatic Hernia
A Short Review
1 in 2000 - 3000 live births

- Failure of complete closure of pleuroperitoneal canal leading to herniation of the abdominal viscera into the thorax, at about 8th week of gestation.
Herniation of abdominal contents:
✓ inhibit normal lung growth
✓ ipsilateral lung hypoplasia
✓ decreased lung weight
✓ decreased number of bronchi, alveoli and in cross section of pulmonary arterial branches

• The hypoplasia may be bilateral
• The degree of hypoplasia is related to the time of gestation when herniation occurred
Three types of CDH:

- Posterolateral Bohdaleck-type (approximately 70%)
- Anterior Morgagni-type (approximately 27%)
- Central septum transversum-type (approximately 2-3%)
Fetal development of the diaphragm: (A) Fusion of pleuroperitoneal membrane, (B) embryological origin of different parts of the diaphragm, (C) central tendon obliterating pleuroperitoneal canal, and (D) fully developed diaphragm
Diagnosis of CDH

Up to two thirds of CDH cases are diagnosed routinely during antenatal US testing in the second or third trimester.

MRI rarely needed to definitively diagnose CDH when US is equivocal; can also assess fetal lung volumes.
Differential Diagnosis

- **Antenatal**
  - Extralobular sequestration
  - Congenital cystic adenomatoid malformation
  - Congenital lobar emphysema
  - Diffuse pulmonary cysts
  - Pulmonary agenesis

- **Postnatal**
  - Pneumothorax
  - True dextrocardia
  - Laryngotracheal obstruction
Prognostic Indicators

- Fetal lung to head ratio
- Liver position
- Left- or right-sided defects
- Large defects
- Patch repair vs. primary repair
- The pressure of the pulmonary circulation
- Coexisting anomalies
- The duration of ECMO
• Cardiovascular anomalies in 23% of patients with CDH
• Increase in pulmonary vascular resistance may lead to right-to-left shunting
• The increase in intra-thoracic components may lead to obstruction of the IVC and decrease in venous return and cardiac output
Signs and symptoms of lung hypoplasia:

- Severe respiratory distress, tachypnea and cyanosis
- Scaphoid abdomen
- Mediastinal contents are shifted
- 5% may present with little respiratory symptoms and may presents with bowel obstruction
Management

- Endotracheal tube
- Nasogastric tube
- Peripheral inserted central catheter (PICC)
- EKG probes
- Chest tube
- Temperature monitor
- Umbilical arterial catheter
- Pulse oximeter (also one on left foot)
Ventilation Strategies

- Intubation immediately after delivery
- Preductal saturation 80% to 95% and a postductal saturation greater than 70%
- Low peak inspiratory pressures (less than 20-25 cm H2O)
- Minimal positive end expiratory pressures (2-5 cm H2O)
- Permissive hypercapnia
- Incorporation of high-frequency oscillation ventilation
Management of Pulmonary Hypertension

- Inhaled nitric oxide (iNO), particularly when there is either an oxygenation index of 20 or higher and/or a preductal–postductal saturation difference of 10% or higher.

- Intravenous prostaglandin E1 or oral sildenafil (a phosphodiesterase inhibitor) may be used.

- No clear benefit has yet been shown in studies conducted to evaluate the efficacy of either iNO or phosphodiesterase inhibitors.
Surgical Management

Surgical candidates are expected to fulfill the following criteria:

• Mean arterial blood pressure is normal for gestational age
• Preductal saturation level is between 85% and 95% on FiO2 <50%
• Serum lactate is <3 mmol/L
• Urine production is >2 mL/kg/h
Advantages of the open repair:

- easier reduction of intra-abdominal viscera
- ability to mobilize the posterior rim of the diaphragm for closure
- easier manipulation of the intestines in cases of rotational abnormalities
- avoidance of thoracotomy-associated musculoskeletal abnormalities
Lap Hernia Repair
Advantages of minimally invasive surgical approaches:

- advantages of smaller incisions
- decreased risk of incisional hernias
- decreased chest deformity
- reduced postoperative pain
- possibly better postoperative pulmonary compliance
Indicators of successful thoracoscopic surgery:

- liver and stomach located in their normal anatomic position
- absence of significant cardiac anomalies
- absence of preoperative ECMO
- peak inspiratory pressure ≤26 cm H2O
- no clinical evidence of PHTN
Disadvantages of thoracoscopic approach:

• higher recurrence rate
• greater operative time and
• the inability to perform a prophylactic Nissen fundoplication for gastroesophageal reflux disease (GERD) in those at high risk
Patch Repair
Various synthetic (polytetrafluoroethylene) and bioprosthetic (porcine small intestine–or dermis-derived matrices) materials have been used in patch repairs.

Synthetic patches have been associated with recurrence rates of up to 50%, which occur in a bimodal fashion as either early (2-7 months) or late (20-51 months).
Permacol: a potential biologic patch alternative in congenital diaphragmatic hernia repair

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Congenital diaphragmatic hernia; Permacol; Recurrence; Gore-Tex

Abstract

Purpose: Recurrence is a well-known complication after patch repair of congenital diaphragmatic hernia (CDH). We propose that a newer, “bioprosthetic” material may lower recurrence rates. The purpose of this study is to compare outcomes of CDH repair with synthetic Gore-Tex (W. L. Gore and Associates, Newark, Del) to the bioprosthetic Permacol (Tissue Science Laboratories Inc, Andover, Mass).

Methods: We performed a retrospective review of 100 consecutive patients with CDH with survival more than 30 days at Children’s Medical Center of Dallas (Dallas, Tex) from 1999 to 2007. The incidence and timing of recurrence, as well as comorbidities were assessed.

Results: Primary repair was performed in 63 patients and patch repair in 37, divided between Gore-Tex (29) and Permacol (8). Overall recurrences were as follows: 1 (2%), 8 (28%), and 0 in the primary, Gore-Tex, and Permacol groups, respectively. Median follow-up was 57 months for Gore-Tex and 20 months for Permacol. Median time to recurrence in the Gore-Tex group was 12 months, with no Permacol recurrences. Both the Gore-Tex and Permacol groups had similar comorbidities, including prematurity, congenital heart disease (76% and 63%, respectively), and the need for extracorporeal membrane oxygenation support (38% and 25%).

Conclusion: Our results suggest that Permacol may have lower recurrence rates compared to Gore-Tex and is a promising alternative biologic graft for CDH repair.

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Fetal endoscopic tracheal occlusion (FETO) has been shown to improve both LHR and pulmonary blood flow. Whether FETO affects survival is still under considerable debate.
<table>
<thead>
<tr>
<th>Table 24-1</th>
<th>Recommended Schedule of Follow-up for Infants with CDH</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Before Discharge</td>
</tr>
<tr>
<td>Weight, length, occipital-frontal circumference</td>
<td>X</td>
</tr>
<tr>
<td>Chest radiograph</td>
<td></td>
</tr>
<tr>
<td>Pulmonary function tests</td>
<td></td>
</tr>
<tr>
<td>Childhood immunizations</td>
<td>Per childhood guidelines</td>
</tr>
<tr>
<td>RSV prophylaxis</td>
<td>RSV season during first 2 years after birth (if evidence of chronic lung disease)</td>
</tr>
<tr>
<td>Echocardiogram and cardiology follow-up</td>
<td>X</td>
</tr>
<tr>
<td>Head CT or MRI</td>
<td>If (1) abnormal finding on head ultrasound examination; (2) seizures/abnormal neurologic findings; or (3) ECMO or patch repair</td>
</tr>
<tr>
<td>Hearing evaluation</td>
<td>Auditory brain-stem evoked response or otoacoustic emissions screen</td>
</tr>
<tr>
<td>Developmental screening evaluation</td>
<td>X</td>
</tr>
<tr>
<td>Neurodevelopmental evaluation</td>
<td>X</td>
</tr>
<tr>
<td>Oral aversion screening</td>
<td>X</td>
</tr>
<tr>
<td>Upper gastrointestinal study, pH probe, and/or gastric scintiscan</td>
<td>Consider for all patients</td>
</tr>
<tr>
<td>Esophagoscopy</td>
<td>X</td>
</tr>
<tr>
<td>Scoliosis and chest wall deformity screening (physical examination, chest radiograph, and/or CT of the chest)</td>
<td>If symptoms</td>
</tr>
</tbody>
</table>

RSV, respiratory syncytial virus; ECMO, extracorporeal membrane oxygenation.
Outcomes of Congenital Diaphragmatic Hernia in the Modern Era of Management

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Objective To identify clinical factors associated with pulmonary hypertension (PH) and mortality in patients with congenital diaphragmatic hernia (CDH).

Study design A prospective cohort of neonates with a diaphragm defect identified at 1 of 7 collaborating medical centers was studied. Echocardiograms were performed at 1 month and 3 months of age and analyzed at a central core by 2 cardiologists independently. Degree of PH and survival were tested for association with clinical variables using Fischer exact test, χ², and regression analysis.

Results Two hundred twenty patients met inclusion criteria. Worse PH measured at 1 month of life was associated with higher mortality. Other factors associated with mortality were need for extracorporeal membrane oxygenation, patients inborn at the treating center, and patients with a prenatal diagnosis of CDH. Interestingly, patients with right sided CDH did not have worse outcomes.

Conclusions Severity of PH is associated with mortality in CDH. Other factors associated with mortality were birth weight, gestational age at birth, inborn status, and need for extracorporeal membrane oxygenation. (J Pediatr 2013; □: □ - □).
References

Conclusions

• CDH is a rare congenital defect associated with poor postnatal outcomes

• A recent transition in ventilation strategies has helped improve survival

• Surgical correction with delayed repair remains the gold standard,

• Fetal intervention is a rapidly developing field that may result in improved outcomes
Thank you!