CONGENITAL DIAPHRAGMATIC HERNIA
Multidisciplinary Management

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Congenital Diaphragmatic Hernia (CDH) Embryology

- Pleuroperitoneal cavity begins in the fetus as a single compartment
- During 5-10th week of gestation, the gut is extruded into the extraembryonic coelom
- Development and closure of the diaphragm is usually completed by 9th week of gestation
- Left side of diaphragm closes later than the right side. Incidence of left CDH is about 80%
CDH - Overview

• Idiopathic defect in muscular diaphragm
• 1 in 2000-5000 live births
• Most are Bochdalek hernias (posterolateral)
• 85% left, 15% right (worse prognosis)
• 60% male, 40% female
Morbidity and Mortality in CDH

- **Lung Hypoplasia**
  - Decreased number of alveoli and terminal bronchioles
  - Decreased type II pneumocytes and surfactant
  - Bilateral - more severe on side of CDH

- **Pulmonary Vascular Hypertension**
  - Excessive muscularization of pulmonary vessels
  - Decreased number of pulmonary vessels
  - Contributes to hypoxia, shunt, and RV Failure

- Iatrogenic
  - Barotrauma
Associated Congenital Anomalies

- 25%-50% associated congenital anomalies
  - Cardiac - PDA, left heart hypoplasia
  - Renal
  - Neurologic - myelomeningocele

- May contribute to mortality
Congenital Diaphragmatic Hernia

Prognosis depends on:
1. Other congenital anomalies e.g., congenital heart disease (15%), renal abnormalities, chromosomal abnormalities, malrotation of the gut, Cantrell’s pentology etc.
2. The degree of hypoplastic lung
3. Persistent pulmonary hypertension of neonate (PPHN)
4. Peri-op management
Antepartum Diagnosis

Vast majority diagnosed by fetal ultrasound
Antepartum Diagnosis

MRI confirmation

fetal stomach

MRI confirmation
Antepartum Diagnosis

Lung-to-head ratio (LHR)
- Measures contralateral lung at 24 to 26 weeks gestation
- <1.0 proposed to identify fetuses at high risk for mortality

Liver herniation
- Proposed to identify fetuses at high risk for mortality

- 15 patients with Lt. CDH, fetal lung-to-head ratio (LHR) was determined by sonography between 24-26 weeks.
- Overall survival rate was 47% with pulmonary insufficiency responsible for all mortality.
- All fetuses with a LHR of less than 1.0 died and all fetuses with a LHR greater than 1.4 survived.
- ECMO was needed for all patients with a LHR < 1.0, 75% of patients with a LHR between 1.0 and 1.4, but only one of four patients with LHR > 1.4.
Antepartum Diagnosis and Management – Fetal Surgery?

- Open fetal repairs no longer performed

- Tracheal occlusion does not improve survival or morbidity in this cohort of fetuses with CDH
CDH – Initial Management

Intra-Partum

- CDH alone is *not* an indication for Caesarian section
- Vaginal delivery preferred if labor/delivery tolerated
- “elective” induction on a weekday morning
  - about 38 weeks
  - minimal maternal sedation
  - consider epidural anesthesia
CDH – Surgical Management

“The Past”

- Immediate/early operation within 48 hours
- Mask or endotracheal anesthesia using cyclopropane or ether plus oxygen
- “vest-over-pants” diaphragm repair
- Aspiration of the ipsilateral chest to “expand” the “compressed” lung
- SURVIVAL POOR – 50% OR LESS
CDH – Medical / Surgical Management
“The Present”

- Delay surgical repair while waiting for pulmonary vascular resistance to fall. It takes around 100 hours on an average. Treat the physiology, not the anatomy!
- minimize barotrauma/oxygen toxicity
- permissive hypercapnea/spontaneous respiration
- no muscle paralysis/alkalosis
- ECMO only for very specific criteria
What is the conventional wisdom regarding delayed surgical repair?

• allows time for remodeling pulmonary vasculature
• delays compliance changes associated with surgery
• salvages infants “too sick” for surgery
• allows ECMO for pre-operative resuscitation
• minimizes need for ECMO

- Moffitt, 1991
- Sakai, 1991
- West, 1992
1) Endotracheal intubation (preferably naso-tracheal intubation for patient comfort and security of tube position.) No “bag and mask.”

2) Gastric tube (double lumen) on continuously low suction. Frequent aspiration of the tube to keep it patent at all times.

3) Umbilical arterial catheterization for blood pressure monitoring and blood sampling.

4) Temperature control, usual supportive treatments

5) Monitoring BP, EKG, Pre- & post-ductal pulse oximetry. Follow pre-ductal $O_2$ saturation (> 90%) to adjust FiO$_2$
6) I.V. fluids:
D/W 10% + calcium to run 4 ml/kg/hr for maintenance
Replacement for gastric tube loss
7) Laboratory work-up:
• Echocardiography if no fetal echocardiogram was done
• Radiograph of chest and abdomen
• ABG’s, CBC, chem 7, blood type & X-match, etc.
Respiratory management: Mechanical ventilation (gentler & kinder).

1) Start with IMV or SIMV, rate 40/min, PIP 20 cmH$_2$O, Ti 0.5 seconds, PEEP 5cmH$_2$O and FiO$_2$ to keep pre-ductal O$_2$ saturation around 90%.

2) Switch to SIMV + PS, or A/C for tachypnea. If rate is not enough to decrease PaCO$_2$, then change to HFPPPV.

3) Try HFPPPV for excessive labored spontaneous breathings or high PaCO$_2$, with rate 100, P 20/1, Ti 0.3 seconds.

4) Try HFO if all 3 above failed. (for pre-ductal O$_2$ saturation<90% or persistent PaCO$_2$>60)

* Most of survivable CDH infants just need (1), (2) or (3).
Congenital Diaphragmatic Hernia in 120 Infants Treated Consecutively With Permissive Hypercapnea/Spontaneous Respiration/Elective Repair

By Judd Boloker, David A. Bateman, Jen-Tien Wung, and Charles J.H. Stolar
New York, New York

- Journal Pediatric Surgery, 2002
- 120 newborns with CDH (1992-2000)
- Permissive hypercapnea, gentle ventilation, delayed repair, no prophylactic chest tube
- 84% survival to discharge

Fig 1. Maximum peak inspiratory pressure required in inborn survivors versus deaths. Survivors (n = 44), Deaths (n = 19). Data not available for 4 deaths.
9) No muscle relaxant. To preserve spontaneous breathing and keep respirator settings low. Mild sedation if needed

10) Positioning patient on ipsilateral side, stabilization, minimal handling, follow ABG’s and correct acidosis

11) Delay surgical repair while waiting for pulmonary vascular resistance to fall (confirmed by echocardiogram, minimal discrepancy between pre- & post-ductal O₂ saturation and high oxygen saturation on low FiO₂.) It takes around 100 hours on an average.
On left: Liver, small intestines and colon in left chest, Stomach and spleen herniated to right chest via retroperitoneal defect

On right: Liver, small intestines and colon in left chest, Stomach shifts after evacuation of air and positioning on left side
12) Vasodilator therapy for severe PPHN (for pre-ductal $O_2$ saturation < 90%,
   • Inhaled nitric oxide
   • Inhaled iloprost (Ventavis, using Aeroneb)
13) Surfactant for the premature infant with RDS and CDH
14) ECMO as a last resort. Surgical repair during or after ECMO
CDH and ECMO - *When is it needed?*

- evidence of sufficient lung parenchyma
  - preductal $\text{SaO}_2 > 90\%$

- when tissue oxygen requirements are not being met by biologic organs and "conventional" care
  - usually manifest by progressive deterioration of oxygenation, metabolic acidosis, and multiple organ failure

- When barotrauma prevents recovery
Intra-operative Management -1

1) Temperature control: heating blanket, Bair Huggar, covers head with plastic bag

2) Monitoring:
   - BP, EKG, end-tidal CO$_2$, ABG’s
   - Pre- & post-ductal pulse oximetry

3) Mechanical ventilation using infant ventilator, no attempt to overexpand hypoplastic lung (hyperinflation of hypoplastic lung causes increase of pulmonary vascular resistanc and is detrimental)

4) I.V. fluids: 4 ml/kg/hr for maintenance, Lactated Ringer’s to replace 3rd space loss and evaporation
Intra-operative Management -2

5) Anesthetic agents depend on oxygenation and cardiovascular stability. No N\textsubscript{2}O
   a) Narcotic (fentanyl 5 ug/kg I.V. bolus, total dose 25 - 50 ug/kg,) muscle relaxant, O\textsubscript{2} or O\textsubscript{2} + air
   b) Low dose inhalation agent, narcotic, muscle relaxant and O\textsubscript{2} or O\textsubscript{2} + air

• For increase of BP and tachycardia, deepen anesthesia.
• For increase of tachycardia and decrease of BP, increase I.V. fluid.
• For suddenly severe hypotension and bradycardia, watch obstruction of inferior vena cava from surgeon's retractor.
6. No prophylactic chest tube to prevent hyperinflation of hypoplastic lung
CDH – Surgical Correction
Open operation - Laparotomy

sub-costal incision
viscera reduced
CDH – Surgical Correction
Open operation - Laparotomy

primary repair

prosthetic repair
CDH – Surgical Correction

What did the lung look like?
Congenital Diaphragmatic Hernia

Time heals all wounds!

39 weeks 4 days old, Pre-op
7 days old, post-op
11 days old
CDH – Surgical Correction
Minimally invasive operation - Thoracoscopic
CDH – Surgical Correction
Minimally invasive operation - Thoracoscopic
CDH – Surgical Correction
Minimally invasive operation - Thoracoscopic
CDH – Surgical Correction
Minimally invasive operation - Thoracoscopic
CDH – Surgical Correction
Minimally invasive operation - Thoracosscopic

- Technical learning curve
- Feasible and safe
- Less pain, less tissue trauma, cosmetically better
- Magnified view for surgeons; better for nurses, trainees and students (except anesthesiologists, PaCO₂ >100 during CO₂ insufflations)
- Early results similar to open repairs
- Long-term follow up needed
CDH – Surgical Correction
Minimally invasive operation - Thoracoscopic

<table>
<thead>
<tr>
<th></th>
<th>“pre-thoracoscopic”</th>
<th>“early thoracoscopic”</th>
<th>“current thoracoscopic”</th>
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<tbody>
<tr>
<td>MIS</td>
<td>Open</td>
<td>MIS</td>
<td>Open</td>
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<tr>
<td>0</td>
<td>208</td>
<td>11</td>
<td>25</td>
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</table>
CDH – Surgical Correction
Minimally invasive operation - Thoracoscopic

Post-op day 1

Post-op day 60
CDH – Surgical Correction
Minimally invasive operation - Thoracoscopic

2 weeks

3 months
Post-operative Management -1

1) Temperature control, usual supportive treatment
2) Monitoring:
BP, EKG
Pre- & post-ductal pulse oximetry and/or TcPO$_2$
3) I.V. fluids: maintenance and replacement of gastric tube drainage
4) Positioning on ipsilateral side dow
5) No muscle relaxant.
Post-operative Management - 2

6) Mechanical ventilation:
IMV, SIMV, Assisted Ventilation, HFPPV or HFV

Avoid hyperinflation of lung.
HFV (for hypoxia or hypercarbia on conventional respirator.)

Aggressive weaning as tolerated

7) Vasodilator continued if needed

8) Morphine 0.1 mg/kg or fentanyl 2-5 ug/kg prn for pain. If > 25ug/kg fentanyl given in operating room, the infant seldom needs analgesic after operation.
9) Positioning patient on ipsilateral side. Mediastinum should gradually shift to ipsilateral side, otherwise, although it is very rare, watch for:

- Pneumothorax on ipsilateral side. Some residual air in ipsilateral pleural cavity after surgical repair is normal.
- Too much pleural effusion requiring partial evacuation (about 40 – 60 ml).
- Hyperinflation of ipsilateral hypoplastic lung. If contralateral lung is compressed and ventilation is compromised, one lung ventilation by block of ipsilateral bronchus may be needed.

10) ECMO is used as last resort.
We have not seen “honeymoon” and post-op ECMO since stopping prophylactic tubal thoracostomy on the ipsilateral side in August 1992.
<table>
<thead>
<tr>
<th>Study</th>
<th>Location</th>
<th>No. Patients</th>
<th>Survival (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Orr &amp; Neff 1953</td>
<td>Kansas</td>
<td>17</td>
<td>53</td>
</tr>
<tr>
<td>Ladd &amp; Gross 1940</td>
<td>Boston</td>
<td>16</td>
<td>56</td>
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<tr>
<td>Donovan 1945</td>
<td>New York</td>
<td>17</td>
<td>76</td>
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<tr>
<td>Gross 1946</td>
<td>Boston</td>
<td>7</td>
<td>100</td>
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<tr>
<td>Harrington 1948</td>
<td>The Mayo Clinic</td>
<td>21</td>
<td>16</td>
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<tr>
<td>Gross 1953</td>
<td>Boston</td>
<td>64</td>
<td>89</td>
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<tr>
<td>Riker 1954</td>
<td>Chicago</td>
<td>31</td>
<td>81</td>
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# Survival in CDH

## 1990s

<table>
<thead>
<tr>
<th>Name</th>
<th>Location</th>
<th>No. Patients</th>
<th>Survival (%)</th>
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<tbody>
<tr>
<td>Reickert</td>
<td>Multicenter</td>
<td>411</td>
<td>69</td>
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<tr>
<td>Ssemakula</td>
<td>Louisville</td>
<td>98</td>
<td>72</td>
</tr>
<tr>
<td>Frenckner</td>
<td>Stockholm, Sweden</td>
<td>52</td>
<td>92</td>
</tr>
<tr>
<td>Wilson</td>
<td>Boston</td>
<td>196</td>
<td>53</td>
</tr>
<tr>
<td>Azarow</td>
<td>Toronto</td>
<td>223</td>
<td>55</td>
</tr>
<tr>
<td>Wung</td>
<td>New York</td>
<td>63</td>
<td>82</td>
</tr>
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# Congenital Diaphragmatic Hernia

**Gentle Ventilation & Delayed Surgery**

<table>
<thead>
<tr>
<th></th>
<th>Immediate Surgery</th>
<th>Very Delayed Surgery</th>
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<tbody>
<tr>
<td>Treatable Infants</td>
<td>17</td>
<td>137</td>
</tr>
<tr>
<td>ECMO Contraindications</td>
<td>2</td>
<td>6</td>
</tr>
<tr>
<td>Non-ECMO Rx’d (%) [survived]</td>
<td>9/15 (60%) [8]</td>
<td>113/131 (86%) [107]</td>
</tr>
<tr>
<td>Overall Survival**</td>
<td>13/17 (76%)</td>
<td>122/131 (93%)</td>
</tr>
<tr>
<td>Survival without ECMO</td>
<td>8/17 (47%)</td>
<td>107/131 (82%)</td>
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** 4 infants discharged on supplemental oxygen**
Conclusions

How has survival increased?

- Prenatal diagnosis allows transfer and delivery at experienced centers
- Advances in supportive care
- Gentle ventilation
- Delayed surgery to allow remolding of pulmonary vasculatures
- No prophylactic chest tube to prevent overexpansion of ipsilateral hypoplastic lung
- Sensible and humane use of ECMO
CDH: Persistent Pulmonary Hypertension

- 49% of pts had normal PAP, PAP/SAP < 0.5, within 3 weeks
  - 100% survived

- 34% had intermediate reductions in PAP, PAP/SAP = 0.5-1.0
  - 75% survival rate

- 17% of pts had persistent PAP/SAP > 1.0 (despite conventional treatment)
  - 100% mortality

Dillon PW et al. J PedSurgery 2004
CDH: Persistent Pulmonary Hypertension

Secondary to a combination of:

- **fixed** elevation of pulmonary vascular resistance secondary to underlying pulmonary hypoplasia (decrease number of pulmonary vessels)
- **reversible** elevation of pulmonary pressure and resistance (excessive muscularization of pulmonary vessels)
CDH: Persistent Pulmonary Hypertension
Columbia Experience

- 2 inborn survivors with CDH requiring ECMO had persistent supra-systemic PAP and depressed RV function and were treated with IV prostacyclin
  - Echocardiograms showed dramatic improvement in the PAP
  - Patients clinically stabilized
  - 2 patient converted to po sildenafil (Viagra) and home

- 1 additional patient receiving po sildenafil for moderate elevation of PAP
CONGENITAL DIAPHRAGMATIC HERNIA

• Global foregut dysmotility

• Gastroesophageal reflux
  – all babies are screened with post-operative pH probe and upper GI series
  – fundoplication reserved for failures of medical management
  – small bowel feeding (jejunostomy) tubes are occasionally indicated
Foregut Ectasia
CONGENITAL DIAPHRAGMATIC HERNIA

Musculoskeletal

scoliosis

pectus excavatum
Long-term management of CDH

- CDH outcomes clinic
- Multidisciplinary – surgery, GI, neonatology, cardiology, pulmonary, and developmental pediatrics, orthopaedics
- Meets once a month
- Convenient for patient follow up as well as outcomes studies
Congenital Diaphragmatic Hernia is Not Necessarily a Grim Diagnosis!

- CDH is a physiologic, not an anatomic emergency.
- Surgical repair can be delayed without fear of reprisal.
- Treat the baby, not the PaO₂ and PaCO₂.
- High airway pressure and FiO₂ can ruin the lungs.
- Iatrogenic disease can be more prevalent than intrinsic disease.
- Most infants with CDH can be saved with a simple ventilator, few drugs, and without the latest device or technique.