Congenital Diaphragmatic Hernia: Historical Perspective and Clinical Update of Post-Natal Management

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The Hospital for Sick Children, Toronto

Robert E. Gross – a pioneer of paediatric surgery

Robert E. Gross

American Journal of Diseases of Children
Volume 76
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Congenital Hernia of the Diaphragm
Robert E. Gross

Congenital hernia of the diaphragm, with some interest, seems to me to have reached a stage where it is usually possible to correct the deformity, regardless of the small size of the subject. It is important to emphasize for the practitioner of paediatric surgery that the operation of choice is the correction of the hernia. The advantages of early correction, in general, are not only evident, but also reflect greatly in the patient's health in a most satisfying manner.

Several recent reviews on the surgical management of congenital diaphragmatic hernia have been published. I should like to place on record the following series of 7 cases of this deformity in which I have personally treated the patients. This small group does not include all of the variations which are not uncommon in congenital hernia of the diaphragm, but it does serve as a foundation for the discussion of the subject. It represents a wide variety of clinical examples which give a picture of the condition and is recommended for children with diaphragmatic hernia. My contact with these young patients has been so full that certain points should be emphasized (or confirmed) which might help in the handling of similar conditions in other communities.

From the Hospital of the Children's Hospital and the Department of Surgery of the Harvard Medical School.

7 cases – all survived!

Congenital diaphragmatic hernia – a surgical problem with a surgical solution

Congenital diaphragmatic hernia

Position or Type of Hernia | Cases | Deaths | Cures
---|---|---|---
Left posterolateral | 53 | 7 | 46
Right posterolateral | 10 | 1 | 9
Esophageal hiatus | 5 | - | 5
Foramen of Morgagni | 4 | - | 4
Total | 72 | 8 | 64

Data from R.E. Gross: Children's Hospital, Boston 1940-1951

The mortality increases
CDH – the non-surgical issues

- Hypoplasia of the ipsilateral and to a lesser extent the good side
- Increased smooth muscle in small pulmonary arteries
- Reduction in cross sectional area of the pulmonary vasculature

CDH – the evolution in management

1940s to 1980s CDH was the quintessential neonatal surgical emergency – urgent repair

- In the 1980s deferred surgical repair and pre-operative stabilisation introduced
- The use of rescue with HFOV and post operative ECMO
- 1990s pre-operative ECMO and HFOV for severe cases
- 2000 – “gentle” ventilation leads to improved survival

Congenital diaphragmatic hernia

Post-natal management

- Deferred surgical repair
- Exogenous surfactant
- Pulmonary vasodilator therapy
- ECMO
- Non-injurious ventilation
- CDH as a cardiopulmonary disease

Deferred surgical repair
Ventilatory Management of CDH
Bohn D. Modern Problems in Pediatrics 1989; 24:76

The use of HFOV in stabilisation of CDH patients

Congenital Diaphragmatic Hernia
Sakai H J Pediatr 1987; 111:432

Deferred surgery – repair makes them worse

Adverse effect on total thoracic compliance associated with repair of CDH

Change in Compliance Following Repair of CDH
CDH 1981-94 - A Tale of Two Cities

The Toronto experience

<table>
<thead>
<tr>
<th>Total patients</th>
<th>pre-operative deaths</th>
<th>post-operative deaths</th>
<th>survivors</th>
<th>hrs to OR</th>
</tr>
</thead>
<tbody>
<tr>
<td>1981-84* 61</td>
<td>6(10%)</td>
<td>23(38%)</td>
<td>32(52%)</td>
<td>9.4(5-31)</td>
</tr>
<tr>
<td>1985-94† 162</td>
<td>56(56%)</td>
<td>17(10%)</td>
<td>89(55%)</td>
<td>40(4-210)</td>
</tr>
</tbody>
</table>

*Urgent surgery era
† Deferred surgery era

Exogenous surfactant in CDH

Surfactant rescue therapy in the lamb model of CDH
O'Toole J Pediatr Surg 1996; 31:1105

Surfactant replacement therapy in CDH
Van Meurs K J Pediatr 2004; 145:312

Term and near-term infants diagnosed prenatally

Survival, need for ECMO & CLD

2284 registry patients
522 diagnosed prenatally
192 received surfactant
Pulmonary vasodilator therapy

iNO in hypoxic respiratory failure in infants with CDH

NINOS Pediatrics 1997; 90:838

Infants with CDH included in a large multicentre RCT of iNO

Infants >34 wks with an OI of >25

Randomised to iNo (25) or control gas (28)

End points were death or the need for ECMO

<table>
<thead>
<tr>
<th></th>
<th>iNo</th>
<th>control</th>
</tr>
</thead>
<tbody>
<tr>
<td>Death</td>
<td>48%</td>
<td>43%</td>
</tr>
<tr>
<td>ECMO</td>
<td>80%</td>
<td>54%</td>
</tr>
</tbody>
</table>

No change in PaO₂ or OI

Echo was not used to diagnose pulmonary hypertension

Pulmonary vasodilator therapy in CDH

iNO has not been demonstrated to improve survival in CDH

An effect on RV pressure can be measured when a TR jet is present, or inferred by change in the direction of ductal shunt or septal curvature

Sildenafil has been beneficial in some patients with CDH and sustained pulmonary hypertension
The use of ECMO in CDH

There is extensive experience with the use of rescue ECMO in both pre and post repair.

Many centres with previously poor results have seen a reduction in mortality.

The overall survival of 5,700 CDH patients in the ELSO registry is 51%.

Morbidity in survivors is high.

Congenital Diaphragmatic Hernia

Long-term outcome in CDH patients following ECMO

Shekerdemian L. J Pediatr 2004; 144:309

- Neurodevelopmental problems in 7 survivors
- Respiratory long-term morbidity 13/27
Does the use of ECMO improve the outcome in CDH?

Congenital diaphragmatic hernia - a tale of two cities

Outcome in newborn infants with CDH 1981-1994

<table>
<thead>
<tr>
<th>Year</th>
<th>Total</th>
<th>Survivors</th>
<th>Percent survival</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Toronto</td>
<td>Boston</td>
<td></td>
</tr>
<tr>
<td>81-84</td>
<td>223</td>
<td>196</td>
<td></td>
</tr>
<tr>
<td>84-87</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>87-91</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>91-94</td>
<td></td>
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<td></td>
</tr>
</tbody>
</table>

Boston management
- 1981-84 no ECMO
- 1984-87 post op ECMO
- 1987-91 pre op ECMO
- 1991-94 permissive hypercapnia

Toronto management
- Rescue therapy with HFOV

Does the use of ECMO improve the outcome in CDH (in an era of gentle ventilation)?
Lung protective ventilation strategies

Pulmonary barotrauma in CDH
Sakuri Y J Pediatr Surg 1999; 34:1813

<table>
<thead>
<tr>
<th>Pathological finding</th>
<th>Bilateral</th>
<th>Ipsilateral</th>
<th>Contralateral</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hyaline membranes</td>
<td>77%</td>
<td>13%</td>
<td>1%</td>
</tr>
<tr>
<td>Pneumothorax</td>
<td>13%</td>
<td>15%</td>
<td>37%</td>
</tr>
<tr>
<td>Interstitial fibrosis</td>
<td>5%</td>
<td>1%</td>
<td></td>
</tr>
<tr>
<td>Parenchymal haemorrhage</td>
<td>46%</td>
<td>4%</td>
<td>50%</td>
</tr>
</tbody>
</table>

N=68

Pulmonary barotrauma in CDH
Sakuri Y J Pediatr Surg 1999; 34:1813

Hyperventilation in PPHN
Drummond WH J Pediatr 1981; 98:603
Management of infants with severe PPHN without hyperventilation

J-T Wung Pediatrics 1985; 76:488

- 15 infants with PPHN
- No paralysis or hyperventilation
- Maximum PIP 25-35 mmHg
- PaCO₂ 40-58 mmHg
- All survived

Outcome in 120 infants with CDH treated with permissive hypercapnia, spontaneous respiration & elective repair


Overall survival 75% (n=120)

<table>
<thead>
<tr>
<th></th>
<th>inborn survivors</th>
<th>inborn deaths</th>
<th>outborn survivors</th>
<th>outborn deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total number</td>
<td>67</td>
<td>23</td>
<td>47</td>
<td>6</td>
</tr>
<tr>
<td>HFOV</td>
<td>6</td>
<td>12</td>
<td>14</td>
<td>5</td>
</tr>
<tr>
<td>HFPPV</td>
<td>38</td>
<td>18</td>
<td>35</td>
<td>5</td>
</tr>
<tr>
<td>ECMO</td>
<td>3</td>
<td>3</td>
<td>7</td>
<td>3</td>
</tr>
</tbody>
</table>

Changing Demographics of Neonatal ECMO

Roy BJ Pediatrics 2000; 106:1334

Congenital Diaphragmatic Hernia

Preoperative stabilisation with HFOV

Miguel D Crit Care Med 1994; 22:S77

- 18 infants with CDH - 11 antenatal diagnosis
- Ventilated from admission with HFOV
- MAP 15 cmH₂O, PIP 30 cmH₂O
- Repaired when MAP 9 cmH₂O, FiO₂ 0.3
- 11/18 survived (72%)
- No ECMO used
### CDH Outcomes HSC Toronto 1995-2001

**N=88**

**N=81**

<table>
<thead>
<tr>
<th>Year</th>
<th>Total</th>
<th>Survivors</th>
<th>ECMO</th>
</tr>
</thead>
<tbody>
<tr>
<td>1995</td>
<td>8</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>1996</td>
<td>8</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>1997</td>
<td>8</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>1998</td>
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<td>2</td>
<td>0</td>
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<td>1999</td>
<td>8</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>2000</td>
<td>8</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>2001</td>
<td>8</td>
<td>2</td>
<td>0</td>
</tr>
</tbody>
</table>

*All patients*  

*Excludes CHD, trisomy 18 and extreme prematurity*

### CDH as a cardio-respiratory disease

All CDH patients have high PVR

Patients with suprasystemic PA pressures that are unresponsive to pulmonary vasodilators will die of RV failure

This can be mitigated by maintaining ductal patency

Cardiac echo is a very important tool in the management of CDH

### Congenital Diaphragmatic Hernia

**HSC Toronto management protocol**

<table>
<thead>
<tr>
<th>Era</th>
<th>Number</th>
<th>Survivors</th>
<th>ECMO</th>
<th>HFV</th>
<th>iNO</th>
<th>PGE1</th>
</tr>
</thead>
<tbody>
<tr>
<td>1981-1991</td>
<td>100</td>
<td>65%</td>
<td>10</td>
<td>20</td>
<td>5</td>
<td>20</td>
</tr>
<tr>
<td>1992-1995</td>
<td>90</td>
<td>77%</td>
<td>15</td>
<td>25</td>
<td>5</td>
<td>25</td>
</tr>
<tr>
<td>1996-1999</td>
<td>80</td>
<td>71%</td>
<td>10</td>
<td>20</td>
<td>5</td>
<td>20</td>
</tr>
<tr>
<td>2000-2003</td>
<td>70</td>
<td>74%</td>
<td>10</td>
<td>20</td>
<td>5</td>
<td>20</td>
</tr>
<tr>
<td>2003-2005</td>
<td>60</td>
<td>97%</td>
<td>10</td>
<td>20</td>
<td>5</td>
<td>20</td>
</tr>
</tbody>
</table>

**n=288**

*“Gentle” ventilation; permissive hypercapnia/hypoxia, use of PGE*

28/29 survivors

- Early (pre-natal) diagnosis
- Use of Apgar scores to identify high risk
- Low PIP ventilation (PIP <25 cmH2O)
- Early use of HFOV (MAP 14-15 cmH2O, PIP 35-40 cmH2O)
**Congenital Diaphragmatic Hernia**

*HSC Toronto management protocol*

- Delayed surgery
- Inhaled nitric oxide may be helpful - use cardiac echo
- Target preductal SaO₂ - no hyperventilation for ductal shunting
- PGE to maintain ductal patency
- ECMO reserved for select cases
- Expect increased morbidity and LOS

**Summary**

Survival in liveborn infants with CDH has improved (80%)

Improved survival is largely due to a change in ventilation practice

The morbidity has increased

The focus should be on managing the cardiopulmonary issues

Infants with CDH should be managed in high volume centres with appropriate expertise

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**Congenital Diaphragmatic Hernia**


**Group 1**  immediate surgical repair with chest drains

**Group 2**  repair delayed for 24 hrs with ECMO rescue, no chest drain

**Group 3**  prolonged pre-op stabilisation with ECMO rescue, no chest drain
**Congenital Diaphragmatic Hernia**


**Ventilator settings**

<table>
<thead>
<tr>
<th></th>
<th>HIPPV</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>PIP</td>
<td>20 cmH₂O</td>
<td>20 cmH₂O</td>
</tr>
<tr>
<td>RR</td>
<td>20 - 40 breaths/min</td>
<td>100/min</td>
</tr>
<tr>
<td>PEEP</td>
<td>5 cmH₂O</td>
<td>0 cmH₂O</td>
</tr>
</tbody>
</table>

**Objectives**

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-ductal SaO₂ &gt;90%</td>
<td>Pre-ductal SaO₂ &gt;80%</td>
</tr>
<tr>
<td>PaCO₂ &lt;60 mmHg</td>
<td></td>
</tr>
</tbody>
</table>

**Groups Comparison**

<table>
<thead>
<tr>
<th></th>
<th>Group 1</th>
<th>Group 2</th>
<th>Group 3</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number</td>
<td>17</td>
<td>28</td>
<td>18</td>
<td>ns</td>
</tr>
<tr>
<td>Weight</td>
<td>3.0</td>
<td>3.1</td>
<td>3.1</td>
<td>ns</td>
</tr>
<tr>
<td>Age at repair (h)</td>
<td>6±6</td>
<td>22±23</td>
<td>100±44</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Survival</td>
<td>14(28%)</td>
<td>21(75%)</td>
<td>17(94%)</td>
<td>p&lt;0.4</td>
</tr>
<tr>
<td>Treated with ECMO</td>
<td>6/17(35%)</td>
<td>7/28(25%)</td>
<td>1/18(6%)</td>
<td>p&lt;0.3</td>
</tr>
<tr>
<td>ECMO contra-indicated</td>
<td>2</td>
<td>3</td>
<td>1</td>
<td></td>
</tr>
</tbody>
</table>

**CDH 1981-94 - A Tale of Two Cities**


<table>
<thead>
<tr>
<th></th>
<th>Survivors</th>
<th>Non-survivors</th>
</tr>
</thead>
<tbody>
<tr>
<td>Time on ventilator (h)</td>
<td>79±154</td>
<td>21.5±70</td>
</tr>
<tr>
<td>HFOV use</td>
<td>13(11%)</td>
<td>74(73%)</td>
</tr>
<tr>
<td>Age at admission (h)</td>
<td>9±14</td>
<td>8.5±28.5</td>
</tr>
<tr>
<td>Pneumothorax</td>
<td>5%(4)</td>
<td>41(41%)</td>
</tr>
</tbody>
</table>