Three Decades of Managing Congenital Diaphragmatic Hernia

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CONGENITAL HERNIA OF THE DIAPHRAGM

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CONGENITAL hernias of the diaphragm occur with such frequency that they can no longer be classed as pathologic curiosities. Surgical attack on these malformations has now 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 or pediatrician that the early recognition and treatment of this deformity can restore the patient to health in a most satisfying manner.

Several excellent treatises on the surgical management of congenital diaphragmatic hernias have previously been published. I should like to place on record the following series of 7 cases of this deformity in which I personally have treated the patients. This small group does not include all of the variations which one can encounter in malformations of the diaphragm, yet the experience is sufficiently broad to serve as a foundation for the discussion of the subject. No personal claim of originality is made for the methods of detecting the condition, for the procedures for surgical correction of the anomaly or for the postoperative care which is recommended for children with diaphragmatic hernia. My contact with these young patients has made me feel that certain points should be emphasized (or condemned) which might help in the handling of similar conditions in other communities.

From the Surgical Service of the Children's Hospital and the Department of Surgery of the Harvard Medical School.
Congenital Diaphragmatic Hernia 1960-80

A surgical problem with a surgical solution
The Effect of surgical repair on respiratory mechanics in CDH

Sakai, H J Pediatr 1987; 111:432

Delayed surgical repair
Congenital Diaphragmatic Hernia 1980-90

Mal-development of the lung and the pulmonary vascular bed

A developmental problem with no simple or urgent surgical solution
Congenital Diaphragmatic Hernia 1980s to 1990s

The era of rescue therapies, pre-operative stabilisation and delayed surgical repair

High frequency oscillation

Extracorporeal membrane oxygenation
Outcome in newborn infants with CDH  1981-1994

<table>
<thead>
<tr>
<th></th>
<th>Toronto</th>
<th>Boston</th>
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<tbody>
<tr>
<td>Total</td>
<td>223</td>
<td>196</td>
</tr>
<tr>
<td>Survivors</td>
<td>122</td>
<td>104</td>
</tr>
<tr>
<td>Percent survival</td>
<td>55%</td>
<td>53%</td>
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CDH 1981-94 - A Tale of Two Cities

Toronto management
Rescue therapy with HFOV
No ECMO use

Boston management
1981-84 no ECMO
1984-87 post op ECMO
1987-91 pre op ECMO
1991-94 permissive hypercapnia
Pulmonary barotrauma in CDH – a clinico-pathological correlation

Sakuri Y J Pediatr Surg 1999; 34:1813

Right (contralateral) lung

Left (ipsilateral) lung

Extensive hyaline membrane formation
### Pulmonary barotrauma in CDH – a clinico-pathological correlation

Sakuri Y J Pediatr Surg 1999; 34:1813

<table>
<thead>
<tr>
<th>Pathological finding</th>
<th>Bilateral</th>
<th>Ipsilateral</th>
<th>Contralateral</th>
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<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></td>
<td>1%</td>
</tr>
<tr>
<td>Parenchymal haemorrhage</td>
<td>46%</td>
<td>4%</td>
<td>50%</td>
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Congenital Diaphragmatic Hernia 2000 onwards

Progressive improvement in survival to >80% associated with changes in ventilation practices

Conventional ventilation with pressure limitation (PIP < 25 mH₂O)

HFOV used with low mean airway pressure (12-14 cmsH₂O)

Frequent use of ECMO with poorly defined selection criteria

Attempts at prenatal intervention
Congenital Diaphragmatic Hernia 2000 onwards

Identifying the severe form of CDH

Data from the CDH Registry (>6,000 patients) shows that birth weight and 5 minute Apgar scores are reliably identify disease severity

These data plus the first post-natal blood gas can be used as ECMO eligibility criteria
Does the use of ECMO improve the outcome in CDH in an era of gentle ventilation?

Overall survival in ELSO registry is 51%

Long term morbidity
  - Neurological deficit
  - Chronic lung disease
  - Gastro-esophageal reflux
  - Musculo-skeletal deformity
Long-term outcome in CDH patients following ECMO: the UK experience

Shekerdemian L J Pediatr 2004; 144:309

n=73

Neurodevelopmental problems in 7 survivors

Respiratory long-term morbidity 13/27
Congenital Diaphragmatic Hernia 2000 onwards

How do we make progress – improve survival and decrease morbidity?
Clinical Commentary

Congenital Diaphragmatic Hernia

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Am J Respir Crit Care Med  2002; 166:911

CDH is a cardiopulmonary disease

Assessment of pulmonary vasodilator therapy by pre/post ductal SaO₂ differences is inaccurate

Need to assess degree of pulmonary hypertension by echo

Shunting at PDA and PFO level

Flattening of IVS

Tricuspid regurgitation

Use PGE₁ to keep the ductus open
Congenital Diaphragmatic Hernia 2000 onwards

Pre-natal intervention
Can we accurately predict a high mortality group of CDH infants prenatally that would justify a foetal intervention?

- Gestational age at diagnosis
- Stomach above or below diaphragm
- PA size by echo
- Liver up
- Ratio of the head to area of right lung
Prenatal intervention in CDH

Deprest J Pediatr Surg 2006; 41:425

Survival to hospital discharge 50% in severe group
The lung to head ratio (LHR) in prenatal prediction of survival in CDH

Deprest J Pediatr Surg 2006; 41:425
CDH – where are we now?

• Early (pre-natal) diagnosis
• Use of Apgar scores to identify high risk patients
• Low PIP ventilation (PIP <25 cmH2O)
• Early use of HFOV (MAP 14-15 cmH2O, PIP 35-40 cmH2O)
• Target preductal SaO2 - no hyperventilation for ductal shunting
• Delayed surgery
• Use cardiac echo to assess PA pressure and status of ductus
• ECMO selection based on excluding patients with severe pulmonary hypoplasia
• Expect increased morbidity and LOS
The future

Will we be able to improve survival to > 80% with prenatal intervention?

Pre-natal intervention needs to be robustly tested in an RCT

The focus needs to be on quality of outcome

CDH patients should be referred to specialised high volume centres
Post-natal definition of severity in CDH


CDH Study Group

1995-1997  n=632

Prediction of survival based on 5 min Apgar and birth weight