New Insights into Managing Congenital Diaphragmatic Hernia

Amir M. Khan, MD
Associate Professor of Pediatrics
UT Houston Medical School
Medical Director NICU, Neonatal Transport and Respiratory Care
Children’s Memorial Hospital, Houston, TX

The speaker has signed a disclosure form and indicated he has no significant financial interest or relationship with the companies or the manufacturer(s) of any commercial product and/or service that will be discussed as part of this presentation.

Session Summary

In this session the speaker will provide a review of the basic pathophysiology of CDH, the current concepts in the management of CDH, as well as a review of the evidence for the management of CDH.

Session Objectives

Upon completion of this presentation, the participant will be able to:

- provide evidence-based preoperative management of patients with CDH;
- review some of the known literature about management of CDH;
- develop an approach to managing patients with CDH.

References


**Session Outline**

See presentation handout on the following pages.
Purpose of the lecture

- To provide and evidence based preoperative management of patients with CDH
- Review some of the known literature about management of CDH
- Disclosure: This is one centers approach at managing patients with CDH

Background information

**CDH**

- Affects 1 in every 3000
- Associated with:
  - Pulmonary hypoplasia
  - Pulmonary hypertension
- Location:
  - 84% on the left
  - 14% on the right
  - 2% are bilateral
- Reported mortality ranges 10-35% (depends on case selection)
- Survival rates have increased over time and are purported to changing management styles from emergent surgery to:
  - Delayed surgery
  - Gentle ventilation
  - Use of ECMO
- Survivors do have higher long term morbidity including CLD and PPHN

1. Lally KJ; Curr Opin Pediatr. 2002; 14: 486-490
3. Lally KJ; Curr Opin Pediatr. 2002; 14: 486-490
CDH

- Standardizing management
  - Makes sense in infrequent disorders such as CDH but does it make a difference

<table>
<thead>
<tr>
<th>Study</th>
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</tbody>
</table>


- Effect of patient volume on outcomes


- An international multicenter randomized controlled trial of high frequency oscillation versus conventional mechanical ventilation in infants with congenital diaphragmatic hernia: the VICI-trial

- Developed a management protocol based on best available evidence. Reiss et al (2010) reviewed the latest data and graded the evidence for each

- We used the protocol as a template and developed a management protocol specific for our patients at CMHH

- Surgery after clinical stabilization and resolution of PHN

<table>
<thead>
<tr>
<th>Variable</th>
<th>Group 1</th>
<th>Group 2</th>
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<tr>
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<td>0%</td>
<td>0.5</td>
</tr>
<tr>
<td>Mortality rate</td>
<td>0.05</td>
<td>0.05</td>
<td>0.5</td>
</tr>
</tbody>
</table>


- Lower peak pressures to avoid barotrauma and volutrauma


Management Protocol
Delivery Room Management

If the diagnosis of CDH is known or suspected:
- Patients should NOT be resuscitated with bag mask ventilation because of gastro-abdominal distention [E].
- Most infants with CDH will have significant respiratory distress at birth and they should be intubated immediately after birth [E].
- Infants with good respiratory effort may be managed according to NRP recommendations.

If the infant is intubated, the infant should be ventilated with an Neo puff with:
- PIP no higher than 22-24 [E]
- PEEP of 5.
- Oxygen should be managed according to NRP recommendations.
- A Replogle tube should be placed in the delivery room to decompress the stomach and after admission to the NICU it should put on suction. [E]

NICU Management

If the infant is intubated in the delivery room, the infant should be placed on the transport ventilator for transport to the NICU. The ventilator should be set for:
- PIP of 22-24 cm
- PEEP of 5 cm
- IMV of 40 b/min
- I-time of 0.35 seconds
- The infant should be monitored in the delivery room and during transport according to NRP recommendations and current NICU practice except:
  - Oxygenation should be assessed ONLY via preductal SaO2.
  - Acceptable preductal SaO2 = 80-95% during the initial stabilization.
  - SaO2 of >70% may be tolerated for several hours to allow enough time for transition.

CDH Management in the NICU

Monitoring
- Similar to all other infants admitted to the NICU except
  - Preoperatively, only preductal pulse oximeter readings should be monitored. [E]

Diagnostic tests
- Similar to all infants admitted to the NICU except
  - An echocardiogram should be obtained to assess cardiac anatomy and the presence or absence of pulmonary hypertension. [E]
  - A head ultrasound should be obtained to evaluate for intracranial hemorrhage prior to starting ECMO. [D]

CDH MANAGEMENT IN THE NICU

- Consults
  - Pediatric surgery
  - Cardiology
  - Genetics

Central lines
- Arterial access: An umbilical arterial line is preferred, but if one cannot be placed, then a peripheral arterial line should be attempted and preferably preductal. [E]
- Central venous access: Either a 2-3 lumen UVC or double lumen percutaneous central line should be placed.

Ventilator management
- Parameter goals for mechanical ventilation [E]
  - Preductal SaO2 of 85-95% as long as oxygen delivery is adequate.
  - During the initial stabilization, SaO2 of >95% may be tolerated for several hours to allow enough time for transition.
  - Postductal (UAC) PaO2: >40 mmHg
  - PaCO2: 50-65 mmHg
  - pH: 7.2 - 7.35
- Adequate oxygen delivery:
  - Lactic acid < 3 (≤ 5 may be tolerated initially)

Van den Hout; Neonatology 2010; 98: 370-380

Admission ventilator settings [E]
- Ventilator type: Dragger or Servo-i
- Mode: Assist Control (Dragger) or Pressure control (Servo-i)
- PIP: 22 cm
- PEEP: 5 cm
- IMV: 40
- I:E ratio: 0.35

Adjust the ventilator to maintain parameters.
- PaCO2 [E]
  - Adjusting PIP or back up rate as needed.
  - PIP should not be higher than 26 and rate should not be higher than 60.
- I:E ratios should be monitored and adjusted as needed to avoid air trapping because of inadvertent PEEP in spontaneously breathing on assist control mode of ventilation.
CDH MANAGEMENT IN THE NICU

- **Change from SIMV to HFOV**
  - The infant should be started on HFOV if
  - Unable to control PaCO2 with a PIP of 26 and a rate of 60
  - Unable to maintain oxygenation with a PIP of 28 and a PEEP of 5
  - Initial HFOV settings should be: [E]
    - Starting MAP should be 2 above conventional ventilator MAP but not higher than 15
    - Starting amplitude may be twice that of MAP or higher to achieve adequate wiggle (wiggle down to the level of umbilicus)
    - Frequency of 8-10 Hz as dictated by the last PaCO2
    - Goal lung expansion for CDH is 8-9 ribs

- **Management of pulmonary hypertension**
  - Pulmonary hypertension may be treated with the measures below if oxygenation does not respond to mechanical ventilation
  - Ensure adequate circulating volume
  - Pulmonary hypertension should be confirmed by echocardiogram

- **Management of PHN**
  - **iNO**
    - No data for its use
    - NiNOS trial showed no benefit and suggested higher need for ECMO or death
    - Most protocol include iNO use but should only be used with adequate inflation

- **Other management of pulmonary hypertension**
  - Inhaled prostacyclin to reduce pulmonary hypertension may be considered in infants who are more than 48 hours old and relatively stable on conventional ventilation with persistent elevated pulmonary pressures [E]
  - Case reports of intravenous prostacyclin in the treatment of PPHN [E]
  - Prostaglandins may be used to unload the right ventricle in the case of a restrictive PDA [E]
  - Such medications should be used after discussion with Pediatric Surgery and Cardiology teams because patients on these medications have to be followed closely with echocardiography

- **Indication for initiating ECMO**
  - Conventional management may be considered a failure and the infant may be placed on ECMO if one or more of the following conditions are met: [E]
    - The inability to maintain preductal saturations above 85% after initiation of HFOV and iNO
    - MAP > 15 mm Hg
    - Oxygenation index consistently ≥ 40
    - Inadequate oxygen delivery with metabolic acidosis defined as lactate ≥ 2mmol/L and pH < 7.20
    - An increase in PaCO2 > 70 resulting in a respiratory acidosis with pH < 7.20, despite optimization of ventilator management
    - Hypotension resistant to fluid therapy and adequate inotropic support, resulting in a urine output <0.5 ml/kg/hour
  - ECMO team should be contacted as per unit routine
### CDH Management in the NICU

- **Weaning Ventilator Support Once Infant is Clinically Stable**
  - Conventional ventilator
    - Wean PIP before back-up rate depending on tidal volume to keep goal PaCO₂.
    - Once the infant is stable on low back-up rate then the mode of ventilation may be changed to SIMV.
    - Adjust FiO₂ as needed to maintain preductal SaO₂ according to goal.

- **Weaning High Frequency Ventilation**
  - Wean FiO₂ to achieve goal preductal saturations.
  - Wean MAP only if the infant is on less than 60% oxygen and lung expansion is at least 8 ribs.
  - Adjust amplitude or frequency to maintain target PaCO₂. Increase frequency before amplitude if frequency is <10.

- **Weaning Inhaled Nitric Oxide**
  - Inhaled NO should be weaned only after the infant is stable on less than 60% oxygen and is on reasonable MAP if on HFOV or PIP if on SIMV. [A]
  - Also discuss the timing of surgery with Pediatric Surgery before weaning pulmonary vasodilators because of a possible increase in pulmonary pressure after surgery.
  - Inhaled NO should be weaned according to unit weaning protocol.

- **Hemodynamic Management**
  - For term and near term infants, the goal MAP should be 40-50mmHg.
  - If initial MAP is <40mmHg
    - Consider normal saline bolus of 10 ml/kg 1-2 times within the first 2 hours. [E]
    - After the initial 1-2 hours, intravascular volume should be carefully assessed and fluid overload should be avoided. [E]
    - If blood pressure cannot be maintained after NS bolus, then consider starting dopamine and dobutamine according to unit practice. [E]

  - Epinephrine may be added as needed, but dose should be no higher than 0.3 microgram/kg/min. [E]
  - Draw serum cortisol level and consider treatment with hydrocortisone, if cortisol level is low or infant is on high doses of pressors or has other clinical indications. [D]
  - Echocardiogram should be obtained in patients who are persistently hypotensive to assess left ventricular function and if LV function is poor than milrinone may be considered. [O]


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Hematological management

- Hematocrit should be maintained according to unit guidelines. However, if the infant is on high oxygen and has elevated lactic acid levels, then hematocrit should be kept >40% [E].
- A DIC screen should be obtained and treated as needed for infants with clinical bleeding or infants with severe acidosis [E].

Antibiotics

- There is no specific indication for antibiotics for infants with CDH [E]

Sedation

- Sedation may be used as clinically indicated but should be used with caution [E]
- Paralysis should generally be avoided and must be initiated with particular caution if the infant is on assist control ventilation [E].

Surgery for CDH

- Surgery may be undertaken after ECMO (if not already repaired on ECMO) or after physiologic stabilization and improvement of pulmonary hypertension [D]
- Infant is considered stable for surgery if: [E]
  - FiO2 < 0.5
  - Mean blood pressure normal for gestational age
  - Lactate < 3 mmol/L
  - No signs of persistent pulmonary hypertension
  - Urine output > 2 ml/kg/hour

CDH Protocol Outcomes

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<tr>
<td>Number of Patients</td>
<td>32</td>
<td>17</td>
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<tr>
<td>Patients treated with ECMO</td>
<td>12 (37.5%)</td>
<td>4 (23.5%)</td>
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<tr>
<td>Overall survival</td>
<td>20 (62.5%)</td>
<td>13 (76.4%)</td>
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<tr>
<td>Survivors in RA</td>
<td>13 (65%)</td>
<td>12 (92.5%)</td>
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