Experience with Cardiac Catheterization in Congenital Diaphragmatic Hernia

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Congenital Diaphragmatic Hernia

- Complex syndrome causing severe hypoxemic respiratory failure
- Associated with high mortality rate
- Affects 1 in 2500-4000 live births
- Associated with
  - Lung vascular and parenchymal hypoplasia
  - Pulmonary hypertension
- Variable post surgical course
CDH Pulmonary Hypertension

- Changes in CDH management have resulted in increased number of survivors
  - Fetal diagnosis / perinatal management
  - Surgical strategies and timing
  - ICU management / ventilatory support (HFOV)
  - Extracorporeal membrane oxygenation (ECMO)
- This has led to increased attention to intermediate and long term outcome for survivors and also complications of therapy and associated abnormalities

Lung Protection and CDH Survival

<table>
<thead>
<tr>
<th>Management strategies</th>
<th>CDH Survival</th>
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<td>Boston n=235</td>
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<tr>
<td>Emergent repair</td>
<td>45%</td>
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<tr>
<td>ECMO/delayed repair</td>
<td>43%</td>
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<tr>
<td>Permissive hypercapnia (PHC)</td>
<td>69%</td>
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<tr>
<td>PHC &amp; lung protection</td>
<td>92%</td>
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Wilson 1997; Azarow 1997; Kays 1999; Bohn 2002; Downward 2003
Slide adapted from R Keller.
Pulmonary Morbidity in CDH
Persistent Respiratory Abnormalities in Long-term Follow-Up Of Congenital Diaphragmatic Hernia Patients: Boston Children’s Hospital


CDH Pulmonary Hypertension

- Pulmonary hypertension continues to be a challenge in CDH cardiovascular management
  - Contributes to prolonged hospitalization and mortality
  - Complicates CDH in majority of patients
  - Long term effects of pulmonary vascular disease in CDH have not been well studied
PH in CDH: catheterization data can be helpful in management

• Clinical significance of PH
  – Determine RA and RV pressure

• Underlying pathophysiology
  – For example, pulmonary vein saturations can help to define cause of hypoxemia vs right to left shunt

• Other contributors?
  – SVC obstruction

• Treatments?

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### Chronic Pulmonary Hypertension in CDH

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<tr>
<th>Patient</th>
<th>Age (yr)</th>
<th>Statuses</th>
<th>PAP (mm Hg)</th>
<th>AOP (mm Hg)</th>
<th>PVRI (UI - m²/PA)</th>
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<td>74</td>
<td>5.19</td>
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Additional Congenital Abnormalities

- Complicate care of 40% of infants with CDH (Cohen et al J Ped 2002)
- Congenital heart lesions account for 2/3
- Chromosomal and other syndromes

Cardiac Catheterization

- Cardiac Catheterization is an important adjunct in CDH cardiovascular management, not just PH
  - Better define physiology and hemodynamics
  - Discover undetected anatomic lesions
  - Provide effective therapy for structural lesions
CDH Patients 2000-2009
CH Boston

- 204 patients, 159 L, 44 R, 1 Bilateral
- Isolated CDH 173
- 48/204 died, survival to discharge 76.5%
- Survival for isolated CDH 82.1%
- Repair performed 188 (92%)
- ECMO 102 (35%)

CDH:
three critical time frames

- Acute – 0-30 days of life
- Late – 30 days of life to first discharge from hospital
- Chronic – Post first hospital discharge (outpatient)

(after Kinsella et al)
Cardiac Catheterization in CDH  
Acute Cohort 1: 0 to 30 days  
2000-2009

- 13 caths, 12 patients
- 9 M, 3 F
- 12 Left CDH
- 12 patch repairs
- 10/12 history of ECMO, 1 x 2, 1 x 3

Cardiac Catheterization in CDH  
Acute Cohort 1: 0 to 30 days  
2000-2009

- Age median 3.2 wks, range 1.9 wks- 4.0 wks
- Associated congenital heart disease: 3 VSD, 1 CoA all pre-repair
- 7/13 caths on ECMO
- 13/13 ventilated, 6/13 iNO
Indications for Catheterization
Group 1

• 5 difficulty weaning ECMO
• 5 Clinical deterioration
• 2 hemodynamics pre cardiac surgery
• 1 hemodynamics

Catheterization Results Group 1

• Baseline Hemodynamics:
  – Mean PAp range 16-61mmHg
  – RA range 2-10mmHg
  – PVR range 1.4-24.3 indexed Woods Units
  – CI range 2.5-4.6 L/min/m2
• 11/12 significant PAH
Catheterization: Structural findings
Group 1

- 2 SVC obstruction (unsuspected)
- 1 CoA (gradient by pre-cath echo)
- 1 LLL Sequestration (unsuspected)

Catheterization Interventions Group 1

- 1 LLL sequestration coil occluded
- 1 coarctation dilated
- 1 SVC balloon dilated
- 1 SVC balloon dilated and stented
Cardiac Catheterization in Pts with CDH – Cohort 2: 30 days to 1st D/C 2000-2009

- 16 cardiac catheterizations
- 12 patients
- 7M /5F
- 10 Left, 2 Right defect
- 12 Patch repair
- 10/12 pts history of ECMO, 1 pt x 2
Cardiac Catheterization in Pts with CDH – Cohort 2: 30 days to 1st D/C 2000-2009

- Age at Cath: median 11.7 wks, range 5.1 wks – 27.1 wks
- 5 pts congenital heart disease
  - VSD x 3, TOF, CoA
  - VSD fenestrated x 2, TOF repair, CoA end to end anastamosis
- 15/16 ventilated, 7/16 iNO

Indications for Cohort 2 Cath

- 6 deteriorating respiratory status
- 4 unable to wean ventilator
- 3 clinical SVC obstruction
- 3 Severe PH by echo
Catheterization Results Group 2

- Baseline hemodynamics
  - RA range 2-22, median 6 mm Hg
  - Mean PAp range 12-68 mm Hg
  - PVR range 2-13.3 indexed Woods units
  - CI 2.1- 6 L/min/m2
- 10/12 significant PAH

Catheterization Structural Findings Group 2

- 6 SVC obstruction (4 unsuspected)
- 1 pulmonary sequestration (unsuspected)
- 1 small PDA
- 1 LPA stenosis
- 1 mild residual CoA
- 1 hemodynamically significant ASD
Catheterization Interventions

- 5 SVC balloon dilation and stent
- 1 SVC balloon dilation
- 1 ASD closure 4 mm Amplatzer
- 1 aortopulmonary collateral coil occluded
- 1 small pda coil occluded
- 1 LPA balloon dilation

Cardiac Catheterization in Pts with CDH – Group 3: Chronic – Outpatient 2000-2009

- 24 cardiac catheterizations
- 17 patients
- 11 M / 6F
- 14 Left, 3 Right defect
- 15 Patch repair, 2 Primary repair
Cardiac Catheterization in Pts with CDH – Group 3: Chronic – Outpatient 2000-2009

- 11 previous ECMO
- Age at Cath: median 5.2 years, range 26.6 wks – 10.2 years
- 6 patients with congenital heart disease (1 repaired TOF, 2 repaired CoA, 1 repaired VSD, 1 ASD, 1 small VSD)
- 15/24 electively ventilated for cath
- 2/24 inhaled NO outpatient at time of cath

Indications for Catheterization Group 3

- 7 suspected SVC obstruction
- 4 increased oxygen requirement
- 13 hemodynamics
Catheterization Results Group 3

• Baseline hemodynamics
  – RA range 3-25mmHg
  – Mean PAp range 13-69 mm Hg
  – PVR range 0.9-26 indexed Woods units
  – CI 2.6-5.4 liters/min/m2
• 10/17 significant PH

Catheterization Structural Findings
Group 3

• 8 SVC obstruction
• 3 LPA stenosis
• 1 large mid-muscular VSD
• 1 large ASD
Catheterization Interventions Group 3

• 1 ASD device placement
• 1 VSD device placement
• 1 LPA dilation
• 1 LPA dilation with stent
• 4 SVC dilation
• 5 SVC dilation with stent

Cath Complications in CDH

• 3 blood loss requiring transfusion
• 2 catheter induced SVT, one requiring cardioversion
• RBBB during catheter manipulation
• Coil embolization, retrieved
• 1 PICC line, 1 Broviac inadvertently removed
**Summary of cath findings in CDH**

- 57 caths in 41 patients
- 28 structural lesions
- 13 Unsuspected structural lesions
- 26 total transcatheter interventions

**Interventions in CDH cath**

- 11 SVC balloon dilation and stent
- 6 SVC balloon dilation
- 2 LPA balloon dilation
- 2 ASD closure
- 2 Coil pulmonary sequestration
- 1 LPA dilation and stent
- 1 CoA dilation
- 1 VSD device closure
Conclusions

• Cardiac catheterization can uncover unsuspected lesions
• Hemodynamics can be used to define physiology
• Transcatheter interventions can provide effective therapy in CDH management
• Low risk of complications even on ECMO at time of catheterization

Important Future Questions

• Define true natural history of late and chronic CDH outcomes
• Characterize prevalence and clinical significance of pulmonary hypertension in late CDH
• Determine optimal imaging modality for late pulmonary vascular disease in CDH
• Define optimal treatments for PAH in late CDH
MRI evaluation of CDH survivors

- Dresden, Germany
- MRI-based pulmonary and cardiac measurements in 12 children operated on for left-sided CDH
- Comparison to age-matched healthy controls
- CDH survivors have:
  - Increased left lung volumes
  - Larger right lung tidal volume
  - Reduced biventricular stroke volume
  - Increased HR and ejection fraction
  - Reduced flow, acceleration time and cross-sectional area of the left pulmonary artery

Abolmaali et al. Eur Radiol 2010 (published online 18 February 2010)
Conclusions

- Pulmonary hypertension (PH) is not uncommon among late survivors of CDH.
- Little is known about the natural history, causes, or treatment options for PH in chronic CDH survivors.
- Because improvements in acute management of CDH are yielding longer term survivors, more research is needed on the late outcomes and health problems of CDH survivors, including PH, and the appropriate treatments.

Chronic PH in CDH

Treatment Strategies

- Encourage growth
- Correct hypoxia if present
- Screen for/correct structural cardiac lesions
- Tx GERD and avoid aspiration
- Prevent pneumonia and URI
- Medical therapy
CT
modified McGoon index = 
\[(RPA + LPA) / \text{descending aorta.}\]
Echocardiographic Predictors of Outcome in Congenital Diaphragmatic Hernia
Hospital for Sick Children, Toronto


SNAP-II Predicts Mortality in CDH

SNAP-II: validated physiologic measure of neonatal illness severity

Lowest BP
Lowest temperature
PO2/FiO2 ratio
Lowest pH
Seizures
Urine output

Traditional Prognostic Factors in CDH

CH Boston Experience 2007-2008

• Most CDH patients have < ½ systemic PAp at 4 weeks of age
• Even elevated PA pressure is consistent with survival, but
• Increased PA pressure is associated with increased length of stay
Needed data

- Larger series of patients from modern multidisciplinary treatment experience
- Detailed analyses of PA pressures and RV function and outcome
- Careful characterization of response to therapy and outcome
- Longer-term follow-up and endpoints beyond survival
Potential Mediators of Increased PVR in CDH

- Insufficient or aberrant eNOS / VEGF expression
- Dysregulation of endothelin (ET-1) and receptor (ET-A, ET-B) function
- Abnormal response in endothelial cells to NO-cGMP pathways to lack of response to vasodilators

VEGF Expression in CDH


(A) Intense expression of VEGF in the medial smooth muscle cells of small diameter pulmonary arteries and in the bronchial epithelium of CDH lung tissue.
(B) Expression of VEGF in the bronchial epithelium and faintly in the smooth muscle cells of the pulmonary arteries of control non-hypoplastic lung tissue.
(C) Expression of VEGF in the arterial endothelium (arrowed) and medial smooth muscle cells of pulmonary arteries in CDH hypoplastic lung tissue.
(D) Arterial endothelium identified (arrow) by CD31 staining using peroxidase technique.
Increased endothelin receptors in CDH patients

Control
Some epithelial expression;
Minimal expression on arterioles

Infant with CDH
Strong expression on arterioles

de Lagausie et al. J Pathol 2005;205:112
Abnormal Expression of Vasoactive Mediators in CDH

• Autopsy series of CDH
  – 23 CDH patients without ECMO, 10 with ECMO
  – 11 controls
• Small pulmonary arteries in CDH patients express less iNOS than controls

• 11 CDH patients, 5 controls
• Decreased heme oxygenase isoform 1 (HO-1) and eNOS in endothelium and arteries in CDH pts

Tyrosine kinase inhibitor:
Targets PDGF-R, c-kit, BCR/abl translocation

**Case report: imatinib in CDH**

- CDH patient with respiratory distress despite NO, sildenafil, and bosentan
- Begins oral imatinib
- Slow improvement over 3-4 months

**Sildenafil for PAH in CDH**

**UCSF Trial**

**Eligibility**

- CDH
- Age 10 to 42 days
- Assisted ventilation and
  - $\text{FiO}_2 \geq 0.40$ at 10-14 d of age, or
  - $\text{FiO}_2 \geq 0.40$ for $\geq 48$ hours at 15-27 d of age, or
  - $\text{FiO}_2 \geq 0.35$ at 28-42 d of age
- or ECMO
- or PA/RV pressure $> 2/3$ systemic
- No known congenital heart disease

**Primary Endpoint**

- N=32
- PVR at 7 week catheterization

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ClinicalTrials.gov NCT00133679
Pulmonary Hypoplasia

- Role of physical compression on development of pulmonary hypertension demonstrated in studies of surgically produced CDH in fetal lambs
- Pulmonary hypoplasia more severe on side of diaphragmatic defect
Dual-Hit Hypothesis Explains Pulmonary Hypoplasia in the Nitrofen Model of Congenital Diaphragmatic Hernia

First hit: genetic and environmental factors

Second hit: interference fetal breathing movements


Genes implicated in CDH in humans or mice

Transcription factors:
- WT1
- FOG3
- GATA34
- COUP-TFII
- Capsulin + MyoR
- MyoD on an mizf background
- RAR-α + RAR-β

Molecules involved in cell migration, mesodermal patterning, or ECM biosynthesis:
- BGF, its receptor c-Met, and the docking protein Grb1
- Slt3 and its receptor Robo1
- Ephrin-B1
- Glypican-3
- Delta-like 3, Lunatic Fringe, and others factors in the Notch signaling pathway
- Elastin
- Type III collagen and other collagen types
- Lysoxidase
- ATP7A
- Fibulin-4
- Fibulin-5

Use of Pulmonary Vasodilator Therapy in CDH
Late PH in CDH

- Cardiac Catheterization may be useful to assess hemodynamics, search for associated cardiac defects and determine optimal therapy

Summary I

- Pulmonary hypertension is a common and potentially a persistent complication in management of CDH
- Limited data on how PAH and management change outcomes in CDH
- Most patients with low pressures at 4 weeks of age; high pressure decreases but does not preclude survival but may increase length of stay.
Summary II

- Little data to show effectiveness of pulmonary vasodilator therapy in management of CDH
- Many possibly effective pulmonary vasodilators are available
- Challenge is in deciding which agents to study and how to study them.

Association of PA Pressure at 4 Weeks and Mean Length of Stay
Outcomes Group 3

- 10/17 significant PH
- 1/24 died
Outcomes Group 1

- 7 of 12 pts died
- 2/7 dead with congenital heart disease

Outcomes Group 2

- 7/12 died
- 4/7 dead with congenital heart disease
Measurement of PA pressures in CDH patients at 4 weeks
Children’s Hospital, Boston

CDH Patients
2007-2008
N=41

Not available for analysis
N=15

Died prior to 4 weeks
0-72 hours, N=6
10-21 days, N=2

Echo at Week 4 +/- 6 days
N=27

CHD
N=4

No Echo @ 4 wks
N=2

PAp < ½ Syst
N=16 (59%)
100% survival

½ Syst < PAp < Syst
N=7 (26%)
86% survival

PAp > Syst
N=4 (15%)
75% survival

Measurement of PA pressures in CDH patients at 4 weeks
Children’s Hospital, Boston

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2007-2008
N=41

Not available for analysis
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86% survival

PAp > Syst
N=4 (15%)
75% survival
PH management strategies in late CDH

- Correct underlying structural cardiac lesions
- Correct hypoxia
- Encourage feeding and growth
- ? Pulmonary vasodilator therapy

Known Changes in Pulmonary Arteries in CDH

Lung biopsy, CDH infant, age 3 months
Non-invasive Delivery of iNO for late PH in CDH
Kinsella et al, J Pediatrics 2003

- 47 infants CDH 1996-2001
- 30 treated with iNO
- 10 with elevated PA pressure with iNO wean, successfully treated with nasal cannula iNO
- Suggests feasibility and physiologic efficacy

Long Term Sequelae in CDH Survivors Post Discharge

- Respiratory issues and chronic hypoxia
- Gastroesophageal reflux
- Chest wall deformity
- Hernia Recurrence
- Neurocognitive delay
- Associated Congenital Anomalies
- Chronic Pulmonary Hypertension
Pulmonary Morbidity

- Pneumonia in 7% of pts with CDH in first year of life (Davis et al, J Peds 2004)
- Preventive strategies include RSV prophylaxis and prompt treatment of URIs
- GERD in 45-90% of infants with CDH
- Early recognition and treatment of GERD to avoid aspiration

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Persistent Respiratory Abnormalities in Long-term Follow-Up Of Congenital Diaphragmatic Hernia Patients: Boston Children's Hospital

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<td>Obstructed PFT with normal V/Q match</td>
<td>1</td>
<td>4</td>
</tr>
</tbody>
</table>

Pts < age 5

Pts ≥ age 5

Chest Wall Deformities

- Pectus deformities and progressive asymmetry of chest wall 21-48%
- Scoliosis 10-27%
Chronic Pulmonary Hypertension in CDH Survivors

- Little known of long term structural pulmonary vascular anomalies in CDH survivors
- Potential for long term pulmonary vascular disease in some patients in both compressed and contralateral lung
Exploration of pulmonary hypertension in early, late and chronic CDH survivors