CDH Treatment and Outcomes: What we’ve learned.

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Director Center for Congenital Diaphragmatic Hernia
Director Extracorporeal Life Support
Johns Hopkins All Children’s Hospital
Background

• Trained in Pediatric Surgery at Columbia University
  – Credit Charlie Stolar
  – Credit Jen Wung
  – Credit Jay Wilson
  – Credit Kevin Lally
  – Thank Matt Harting and the CDH community for asking me to speak
I have no disclosures

- >450 CDH patients
  - 321 at University of Florida
    - 1992 - 2015
  - > 140 patients at Johns Hopkins All Children’s Hospital
    - 2016 - present
Outline

• Describe our population
  – 101 consecutive CDH cases at JHACH
  – Describe them by risk stratifiers
    > Anatomy, lung volumes, physiology, associated anomalies

• Describe the care paradigm
  – Foundational principles
  – Ventilation
  – Focus on ECMO
  – Focus on Repair
Describe Outcomes

• Survival
• Time in hospital
• Outcomes
  – Neuro imaging outcomes (gross)

• Conclusions
This is the disease: Pulmonary Hypoplasia (highly severe)
CDH Referral Pattern

High volume Referral Center
High percentage of prenatally diagnosed and evaluated patients
Increased Severity
Johns Hopkins All Children’s
St Petersburg, FL
Lessons Learned, treatments refined

- >450 CDH patients

- 321 at University of Florida
  - 1992 - 2015

- > 140 patients at Johns Hopkins All Children’s Hospital
  - 2016 - present
Analogy: Golf

• Golf is a HARD game
• To succeed: ALL ASPECTS of your game need to be good
  – Drives
  – Long irons
  – Short irons
  – Chipping
  – Putting
  – Rescue

  – One bad shot can ruin any hole
CDH care is hard.

To succeed at CDH care, it’s not just one thing. There is no single “secret”

5 major lessons learned

-Lungs: the primary key to survival
-Repair: the second key to survival
-ECMO: Critical to save the worst
  -Must do Better ECMO
-Risk stratification: know your patient
-Offer your best therapy to your sickest patients
-Belief: they do have enough lung to survive
Hypothesis:
- Hyperventilation/alkalosis is harmful to CDH patients
- Elimination of this therapy will result in improved survival
- Prospective change in therapy in August, 1992

Kays, Langham, Ledbetter, and Talbert
CDH: Treatment Strategy

- Light to moderate sedation (no paralysis)
- Conventional SIMV pressure-limited ventilation with rate set to patient comfort and clinical state
- Lowest pressure which provides adequate chest movement (usually 20 - 24 cm H2O)
- Hyperventilation and alkalosis are strictly avoided

Indications for ECMO

• Inability to maintain and insure adequate oxygen delivery to the brain
  – Pre-ductal sats < 85%
  – NIRS < 50%
  – Despite optimal support
Mean PIP over 120 hours

Mean +/- SEM

p<0.05 at all time points

p=0.00001

Time*Era effect
Mean PIP over 120 hours

Mean +/- SEM

p<0.05 at all time points

p=0.00001

Time*Era effect

Survival Curve by Era, All Patients

Survival Graph

p<0.0001

Must eliminate any iatrogenic lung injury:

The number of CDH patients that survive is all about how well we take care of their lungs
CDH Treatment Fundamental #2

- (2) Repair the Hernia (CDH) (n=268)
When to repair

• Avoided ECMO:
  – Follow clinical course. When improvement plateaus, repair
  – Day 4 – 7 (mean 118 +/- 27) hrs
Early repair before ECMO vs Delay and arrive to ECMO unrepaired (w/ opportunity)

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Pros and Cons of “Repair before ECMO”

- **Pros**
  - It works. ECMO runs are easier, cleaner, better.
  - Minimal risk of bleeding
  - New comfort going to ECMO.
  - **Everyone gets repaired.**

- **Cons**
  - Repair becomes time sensitive:
  - Still concern could increase risk of ECMO
  
  - BUT WHY ALL THIS EFFORT???
In early 2016, we transitioned from early repair “BEFORE ECMO”, to early repair ON ECMO

- Repair next am
- Ave time to ECMO: 30 hrs (+/- 33)
- Ave time to Repair: 65 hrs (+/- 69)
- Next morning is most common time for repair after initiating ECMO
Principle #3
Do Better ECMO

• Decision making and timing
• Better Circuits
• Better anticoagulation
• Better concepts
  – Support and weaning
Better ECMO:

- All VA. (VV doesn’t unload RV nor PA’s)
- Repair early on ECMO. 24 hours
- Better anticoagulation:
  - Bivalirudin
Bivalirudin

• Direct thrombin inhibitor
• Clean
• Predictable
• Efficacy?
  – Bleeding vs clotting?
• Pharmacokinetics
  – 20% renal excretion
  – 80% proteolytic degradation
    • ? Where ? (important)
Heparin Drip 2500 units in 50ml D5W started.

300 units Heparin.
Anticoagulant Properties (?)

Clot ← Heparin → Bleed

Bivalirudin
ECMO Pumps

• Roller vs Centrifugal?

• Below 10 kg, not all centrifugal are created equal
Offer your best treatment to your sickest patients. Believe they can survive.

- What are the outcomes in "the worst" CDH patients?
  - (Buckets A&B)

Kays DW, Islam S, Perkins JM, Larson SD, Taylor JA, Talbert JL

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**Most Severe 10%: (N=172) Survival 8/17 = 47%.**
Survival vs Time on ECMO

Proportion Surviving, All Patients (With 95% CI For Each Day)

J American College of Surgeons, 2014
Kays, Islam, Larson, Perkins, Talbert
Association of risk factors with Duration

- Estimated that ECMO duration increase by 2.0 days for each unit decrease in Apgar-1
  \[ p < 0.001 \]

- Estimated that ECMO duration increases by 2.1 days for each 1 unit decrease in Apgar-5
  \[ p < 0.001 \]

- Estimated that ECMO duration increases by 0.2 days for each unit decrease in Predicted Survival
  \[ p < 0.001 \]

- Estimated that ECMO duration increases by 0.8 days for each 0.1 unit drop in LHR
  \[ p = 0.006 \]
1st Run ECMO vs 2nd Run ECMO for CDH

1st Run ECMO

2nd Run ECMO
What if we put it all together?

- Protect lungs
- Risk Stratify Repair timing
- Get Everyone Repaired
- Do Great ECMO
  - Good decision making
  - Minimize errors
- Believe they can Survive
- What If?
CDH Program @ JHACH

- 101 Consecutive patients
- Unselected. All-comers*
  - *2 patients seen at our program chose to deliver at their home hospital. Both FDIU
    - Bilateral CDH with 2% o/e TFLV
    - Trisomy 15 mosaic with hydrops
Our Paradigm

• CDH is about lung hypoplasia
  
  – All treatment decisions are about gas exchange and about helping little lungs work as well as they can.

  – Pulmonary Hypertension is a secondary issue, and does not drive management
Treatment Specifics

- Prenatally evaluation including
  - LHR, Echo, and MRI (o/e TFLV)
  - Counseling
- Inborn Delivery at 38 weeks or so
- Resuscitation in Delivery Room by CDH Team
  - CDH surgeon, CDH neonatologist, CDH RT, CDH nurses
  - (Roles meld and titles fade)
- Conventional ventilation,
  - PIP 25 or less
  - Pre-ductal sats most important
  - Nitric Oxide started for near ECMO level hypoxemia
    - Pre-ductal sats less than 85, PO2 less than 35
  - ECMO when unable to maintain pre-ductal sats at or near 80 - 85 despite optimization of support (brain protection)
Treatment Paradigm

• Risk stratify repair timing to minimize risk of ECMO
• Delay repair for 4 – 6 days (as long as improving)
• If goes to ECMO, repair within 24 hrs
  – Pediatric specific centrifugal or rollerhead pump
  – Bivalirudin probably better than heparin
  – Do GREAT ECMO: good decisions, good supportive care, time
  – Develop exceptional surgical technique and expertise

• Focus on lung function and gas exchange
  – Pulmonary hypertension is the symptom, not the disease

• Believe they can survive
  – Minimize Errors
  – Learn from mistakes
  – Simplify care
ECMO Management

• VA ECMO

• Pump:
  – Sorin Revolution at JHACH (3 patients then changed)
  – Pedi-Mag for all subsequent ECMO (14 cc prime)

• Anticoagulation
  – Changed to Bivalirudin (3/1/2016)
ECMO Weaning

• Athletic Training Paradigm

  – Wean ECMO at a (slow) rate that allows the heart and pulmonary vasculature to develop work capacity over time.

  – All ECMO patients started on sildenafil at 0.8 mg/kg/d when start wean phase (to help stabilize pulm vasc)

  – All patients successfully weaned and none required a second ECMO run.
Second Axis of Severity: CDH Groups (Buckets)

The full spectrum of CDH:
- Associated anomalies: None
- "Isolated CDH"

The full spectrum of CDH:
- Associated anomalies: less severe, not life threatening
  - ie. Small to moderate VSD, partial renal obstruction
  - less severe genetic defects

The full spectrum of CDH:
- Associated anomalies: severe to life-threatening
  - major chromosomal (trisomy 13, 15, 18, others)
  - major heart defects. (STAT 3 or higher?)
    - single ventricle physiology (HLHS, pulm atresia-VSD)
  - bilateral CDH
  - major abd wall defect: Giant Omphalocele
  - major CNS anomaly
101 Consecutive patients

- Bucket A
  - (Isolated)
  - 71
- Full spectrum of disease

- Bucket B
  - Assoc. Anomalies
  - 20
- Large VSD: 2
- DiGeorge Syndrome
- Neonatal Diabetes
- Kleinfelter
- Obstructive Uropathy
- Serious but non-lethal chromosomal abnormalities

- Bucket C
  - Severe Assoc
  - 10
- Bilat CDH-2
  - TFLV 6% and 8%
- Complex Card-4
  - Single vent
  - Pulm atresia/VSD
  - TA w/ IAA
  - TAPVR w/ Em. Syn
- Giant Omph.-2
- Massive hydrocephalus
JHACH Patient Distribution

**Defect Anatomy**

- **Left (Up):** 57
- **Left (Down):** 26
- **Right:** 16
- **Bilateral:** 2

**Buckets**

- **A:** 71
- **B:** 20
- **C:** 10
58 of 101 had TFLV o/e less than 30%. (58%)
# 9 worst patients by 1 hour ABG

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Figure 1: Distribution by Severity by Defect Size

A B C D

JHACH CDH Registry

Frequency %
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<td>1.8 (0.8)</td>
<td></td>
</tr>
<tr>
<td>Risk Stratifier</td>
<td>All CDH (n=101) Mean (SD)</td>
<td>No ECMO (n=37) Mean (SD)</td>
<td>ECMO (n=66) Mean (SD)</td>
</tr>
<tr>
<td>-------------------------</td>
<td>----------------------------</td>
<td>--------------------------</td>
<td>-----------------------</td>
</tr>
<tr>
<td>APGAR 1 min</td>
<td>3.35 (2)</td>
<td>4.8 (2)</td>
<td>2.5 (1.5)</td>
</tr>
<tr>
<td>APGAR 5min</td>
<td>5.94 (2)</td>
<td>7.1 (1.7)</td>
<td>5.2 (1.8)</td>
</tr>
<tr>
<td>CDH SG Predicted Survival</td>
<td>60.7 (21)</td>
<td>76.5 (12)</td>
<td>51.6 (19.8)</td>
</tr>
<tr>
<td>LHR</td>
<td>1.06 (0.4)</td>
<td>1.36 (0.54)</td>
<td>0.93 (0.28)</td>
</tr>
<tr>
<td>o/e LHR</td>
<td>36 (15)</td>
<td>47 (18)</td>
<td>31 (10)</td>
</tr>
<tr>
<td>MRI-1 TFLV o/e</td>
<td>27 (13)</td>
<td>40.4 (12)</td>
<td>22 (8)</td>
</tr>
<tr>
<td>MRI-2 TFLV o/e</td>
<td>24.5 (9)</td>
<td>28.8 (6.8)</td>
<td>23 (9)</td>
</tr>
<tr>
<td>PH</td>
<td>7.07 (0.19)</td>
<td>7.24 (0.13)</td>
<td>6.97 (0.14)</td>
</tr>
<tr>
<td>PCO2</td>
<td>91 (36)</td>
<td>61.5 (24)</td>
<td>108 (30)</td>
</tr>
<tr>
<td>PO2</td>
<td>77 (101)</td>
<td>131 (146)</td>
<td>45 (32)</td>
</tr>
<tr>
<td>Lactate</td>
<td>3.3 (3.65)</td>
<td>1.8 (0.8)</td>
<td>4.0 (4.2)</td>
</tr>
</tbody>
</table>
ECMO Survival to D/C

CDH-ECMO Anatomy

- Left CDH: 51 Placed, 49 Survived
- Right CDH: 11 Placed, 11 Survived
- Bilateral CDH: 2 Placed, 0 Survived

Buckets

- Bucket A: 43 Placed, 42 Survived
- Bucket B: 13 Placed, 13 Survived
- Bucket C: 8 Placed, 5 Survived
JHACH CDH Survival

**Overall CDH Survival Rate**
97/101 = 96%

**CDH without Major Associated Anomalies**
90/91 = 99%
JHACH MRI TFLV observed to expected (All Buckets)

- TFLV o/e 20-30%: 22
- TFLV o/e 16-20%: 22
- TFLV o/e 11-15%: 11
- TFLV o/e < 10%: 3

Survived:
- TFLV o/e 20-30%: 21
- TFLV o/e 16-20%: 22
- TFLV o/e 11-15%: 11
- TFLV o/e < 10%: 0
Time in Hospital

- No ECMO
  - Extubation: 12.7 (+/- 6 days)
  - Discharge: 1.5 mos (+/- 0.9)

- ECMO
  - Extubation: 32 (+/- 33) days
  - Discharge: 2.42 (+/- 2.2) mos

95/97 went home breathing spontaneously

2 tracheostomies, both from Bucket C
What we’ve learned

- Focus on the lungs
- Repair the CDH
- Do exceptional ECMO
- Believe they can survive
What we’ve learned

• Pulmonary hypoplasia in CDH needs not be lethal
• We currently have the tools necessary for exceptional outcomes.
• Survival in CDH without major associated anomalies can approach 100%
• We can look prenatal patients in the eye and quote 95% predicted survival
CDH

- Quantity of Survival
  - Care of lungs

- Quality of Survival
  - Care of brain

- (Another talk)