Outcomes of CDH Patients

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Session Summary
How do CDH kids do long-term? What are typical outcomes, and how do outcomes vary across the CDH spectrum, and in different centers? What should the astute clinician look for in the short term and in the longer term?

Session Objectives
At the conclusion of this activity, participants will be able to:

- discuss how the spectrum of CDH severity relates to the risk of long term morbidity in CDH;
- describe the six major areas of focus for follow-up issues in CDH survivors;
- describe the range of neurocognitive outcomes in CDH survivors.

References


Follow-Up Issues: Congenital Diaphragmatic Hernia

He survived, but how is he??

CDH is a Spectrum of Severity
Definable by

- Anatomy (before)
  - Left liver-down
  - Left liver-up
  - Right
- Physiology (during)
  - Apgars
  - Blood gases
- Therapy required (after)
  - Patch/ No Patch
  - ECMO/ No ECMO

Spectrum of Severity affects Spectrum of Outcomes

- Less Severe CDH, less morbidity
- Quality of care affect outcomes too:
  - Better center, better results

Problem with CDH f/u literature
(and CDH literature in general.....)

- It is (mostly) NOT risk stratified
- Conclusions are "CDH patients ....." 
- The CDH spectrum of severity is WIDE
- Interpret all CDH papers and f/u studies with this knowledge.

General Overview

- Patients with stomach down L CDH
  - Should lead normal lives with minimal morbidity
- Patients with stomach up, liver down CDH
  - Should lead normal lives with minimal morbidity"
- Patients who survive left liver-up (LLU) CDH
  - Are at risk for the full spectrum of morbidity
- Survivors of Right CDH are a spectrum of their own
  - Higher risk of persistent PH.
6 areas of follow-up

- Lung function
- Heart function
- GI & nutrition
- Musculoskeletal issues
- Diaphragm Integrity
- Neurodevelopmental Outcomes

A small lung

Discharge and Respiratory Outcomes

- Average time to discharge for survivors of worst 10%:
  - 3.3 months (1.6 – 4.2)
- No patients required tracheostomies or home ventilation.
  - Discharged home on nasal cannula oxygen.

Newborn from earlier slides pH 6.64,...

Lung Function. Check post-ductal sats

- Outcomes related to
  - degree of pulmonary hypoplasia
  - degree of lung induced lung injury from ventilatory support
- Less Severe patients (LLD and less severe right)
  - Usually don’t require O2 at d/c
  - Should consider synagis year 1 (yes if ECMO or O2 at discharge)
  - May benefit from inhaled steroids and bronchodilators if intermittently symptomatic
    (asthma-like symptoms)
- More to Most Severe
  - Usually on O2 at discharge, off by several months
  - May be re-admitted several times first year for respiratory illness. Resolves
  - May benefit from inhaled steroids and bronchodilators (often receive)
  - Get better over time.
- All
  - May have less endurance and stamina, but
  - Benefit from physical activity
  - Encourage sports
  - Can run a marathon (but won’t win!)

Pulmonary Follow-up

- Measurable deficits but are they clinically relevant?
- 72% with normal PFT’s
- Normal lung volumes, decreased flow,
- 28% with evidence of obstructive airway dz

Heart

- At risk for anatomic abnormalities
  - ASD, VSD, PDA, (and more complicated)
- At risk for chronic changes from unresolved PHTN **
  - Spectrum of severity
    - Follow-up echo’s
      - At 1 – 2 years
      - At 5 – 6 years
      - At 10 –12 years
- Cardiac flu does not need to be intense, just someone has to do it
- (Completely unclear if medications commonly used for chronic pulmonary hypertension. i.e. sildenafil, tadalafil, flolan, etc.)

Muratore et al J Ped Surg 2001
GI (GERD) & Nutrition

- Again, the spectrum of severity
- Virtually all have increased GER
  - Abnormal diaphragm
  - Vertical stomach
  - Breathe harder
- 75% CDH survivors have reflux
- Worse CDH, worse reflux
- 25 – 40% undergo anti-reflux procedure
- Reflux decreases with age
- 38% still have pathologic reflux at age 5 (1)
- Long term risk of esophageal disease: unknown

Canuso, AM et al.
GER in CDH Patients Ped Surg Int 2013

GERD w/u

- UGI is a reasonable first start
- They WILL be malrotated (part of CDH)
- Severe reflux needs surgical Rx
  - Lung complications
  - Inability to gain adequate weight

Growth & Nutrition

- How much is enough?
  - 56% are below the 25th percentile
- What are downsides of G-tube??
  - Depression of oral feeds. Oral aversion
- Our severe patients are eating PO,
  - are below the 5th percentile,
  - are following curve
  - are developing well
- We avoid g-tubes, and transition away asap

Skeletal Deformities

- Pectus Excavatum: 47% (with patch repair)
- Scoliosis

Kuklova P. Ped Surg Int 2011

Very large Goretex patch repair

Diaphragm Integrity

- CXR yearly
- Rate of recurrence is related to:
  - Size of defect (patch/no patch)
  - Surgical technique
  - Recurrence rates 5 – 40% (at 1 year!)
  - Most recurrences are not an emergency to repair
  - Most recurrences are w/o specific symptoms
CDH

• Quantity of Survival
  is directly related to how well we take care of their LUNGS

Quality of Survival

• Directly related to:
  • How well we take care of their brains

Neurodevelopmental outcome

• At 29 months:
  – 77% are normal/23% are delayed
• Results are likely center specific
• Risk of both global and focal deficits
• Better brain monitoring may lead to better outcomes

CDH Survivor

Focused Protocols

• Neuro-protective
• Brain monitoring
  – NIRS
  – Biomarkers of brain injury
  – Correlation with Outcomes
NIRS Brain Sat monitor

Liver-Up CDH
Diagnosed prenatally at 14 weeks
LHR 0.69
At birth, on VA ECMO w/in 4 hours
Repairs on ECMO at 3 days
2 ECMO runs lasting 29 days

Putting a face with the numbers

NP

Passionate people and families

Conclusions

• CDH is a serious disease but survival in the best centers exceeds 85%
• The less severe half of the spectrum is totally normal
• The more severe half may deal with issues including affecting growth, skeleton, pulmonary, GER, and neurodevelopment.
• Most issues resolve in the first few years
• Overall outcomes excellent
• Long term issues:
  – related to GER may arise later
  – malnutrition, bowel obstruction
  – neurodevelopmental

Thank You:

Johns Hopkins All Children’s
Johns Hopkins School of Medicine
University of Florida

Research and Program Coordinator
Joy Perkins, RN, RRT

Partners and Mentors
Charles Stolar, MD  Jen T. Wang, MD
Max R Langham, MD  Paul Danielsen, MD
James L. Talbott, MD  Paul Colombani, MD
Saleem Islam, MD  Shawn Larson, MD
Nicole Chandler, MD  Janice Taylor, MD
David Burchfield, MD  Mike Weiss, MD
Johns Hopkins All Children’s

Pediatric Surgery Fellows
Dan Neal, Statistician