CONGENITAL DIAPHRAGMATIC HERNIA

I’M SOOO NERVOUS!!
I FEEL LIKE MY STOMACH IS IN MY CHEST!!

presented by

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PRACTICE GAP: Lack of knowledge and management of babies with CDH.

DESIRED OUTCOME: Through a multi-disciplinary review on outcomes of babies with Congenital Diaphragmatic Hernia delivered at our facility, increase awareness of modern management of CDH and its implications.
I have no financial or non financial disclosures to disclose today
C. Everett Koop was 13th Surgeon General of the United States under Reagan.
BACKGROUND

• Seen in approximately 2-4/10000 pregnancies
• Generally a sporadic event with very rare hereditary causes
• Often times associated with other abnormalities
  • Aneuploidy
  • Syndromes
  • Other midline defects
  • Cardiac (about 10% which had lower survival)
• Survival rate influenced by the presence of other anomalies
Fig. 1. Congenital Diaphragmatic Hernia Study Group overall mortality by year.
WHAT ABOUT THE DIAPHRAGM

• It’s pretty darn important!!!! (Remember Voicebox on 8/1)
• Develops early on and separates the abdominal contents from the thoracic stuff
• Lack of barrier allows abdominal contents to enter chest resulting in multiple issues
• Biggest issues
  • Pulmonary hypoplasia
  • Pulmonary hypertension
CDH SPECIFICS

- **Location**
  - 95% are posterolateral (Bochdalek) More russian influence?????
  - Can occur in other locations
- 80+% are left sided
- 15% are right sided
- Can be posterior or at/near where structures transverse through the diaphragm
Pau Gasol 2010 Game 7 victory over Celtics
ULTRASOUND
STUFF
YAY!
YAY!
YAY!
4CH BBY BREECH
PREDICTORS OF OUTCOME

• Right sided CDH
• Liver in chest on left sided
• Polyhydramnios
• Other associated anomalies
• Stomach in the chest
PREDICTORS OF OUTCOME

• It’s all about the lungs!!!!!
• Important to know the amount of tissue and maybe function???
• Various things
  • LHR (mostly use observed to expected)
  • Lung volume (both 3D US and MRI)
  • Estimate degree of pulmonary hypertension
  • Oxygen saturation testing
  • Composite scoring system
LUNG HEAD RATIO

• Can be just the LHR
  • If LHR < 1.0  poor prognosis
  • If LHR 1.0 – 1.4 often times need ECMO
  • If LHR > 1.4  better prognosis

• Observed/expected LHR (lungs grow 4x faster than head)
  • < 0.15  extreme severity
  • 0.15 -0.25 severe
  • 0.26-0.35 moderate
  • > 0.36 mild

Only accurate sometime after 23 weeks, really probably 28-32 weeks
PREDICTORS

- All infants dx’d with left sided CDH from Jan 2002 to June 2005
- Presence or absence of liver along with LHR
- Evaluated survival to discharge and need for ECMO
- Looked at outcomes with LHR < 1.0 or > 1.0
- 28 infants met criteria
- Overall survival was 24/28 infants (86%)

PREDICTORS

• Survival based on LHR
  - ≤ 1.0 8/11 (73%)
  - > 1.0 16/17 (94%)  p = 0.11

• Need for ECMO
  - ≤ 1.0 3/11 (27%)
  - > 1.0 1/17 (6%)  p = 0.11

• Outcomes based on liver position
  - Survival if up 9/12 (75%)
  - Survival if down 15/16 (94%)  p = .16
  - ECMO if up 3/12 (25%)  p = .16
  - ECMO if down 1/16 (6%)

(no difference in LHR between 2 groups)

 Arkovitz MS et al, J Ped Surg, 2007; 42:107-111
Timothy Crombleholme, MD (Cincinnati, OH): While I would readily concede the LHR is not perfect, and one of its biggest challenges is actually doing it correctly, having been involved with now 2 startup fetal therapy programs, both in Philadelphia and in Cincinnati, I can tell you from firsthand experience how difficult it is to get even really superb ultrasonographers to get an accurate LHR. We had values all over the map when we started in Philadelphia and the same was true in Cincinnati. So one important question is, did you have your sonographers, who I know are superb at Columbia having worked with them—were their measurements validated by someone with experience in measuring LHRs? That’s number one.

Number two, the LHR has only been proven to be valid when it’s obtained between 23 and 26 weeks. You had patients who were having them measured at 36 weeks, and we’ve looked at that, completely unreliable.

Marc S. Arkovitz, MD (response): I’m going to address the questions backward. As far as the liver herniation, we did not by ultrasound in any way quantitate the liver herniation, so I can’t answer that question because I don’t really know. It was just whether or not there was evidence of fetal liver herniation into the ipsilateral hemithorax.

The gestational age is a very valid criticism but we did actually perform a regression analysis and what we found was that the gestational age seemed to correlate with the fetal LHR ratio so that as you got longer into gestation the LHR seemed to increase. I think if we can develop a large prospective study that we do have to normalize measurements all to be included in the time period that you mentioned.

As far as the ultrasonographers being—I guess they would have to go to San Francisco to be trained or ???
Hanmin(?) Lee, MD (San Francisco, CA): Just to echo Tim’s comment about the difficulty of measuring LHR, it’s a very difficult number to measure. One would think that if the ultrasonographer gives you a value of 0.9 that that’s written in stone but in fact there is a range of LHRs. We use the lower range as the number that we use, so if you have an LHR of 0.8 to 1.0, we use the 0.8 number. The other thing is that there are many different ways actually that people measuring LHR and they have interpreted the initial papers very differently, and so it is actually a very difficult thing to measure. There is a lot of center to center variability in the measurement of LHR. The other point is that we’ve looked at predictability over gestational ages and before 24 weeks the predictability goes way down. It was initially described I think between 24 and 26 weeks, but we’ve actually found that at least in our series of several hundred that actually the best predictability comes later in gestation from 26 to 32 weeks.

Marc S. Arkovitz, MD (response): Thank you. I think I’ve already made the point about doing a multicenter randomized prospective study has already been addressed. I guess I would just say that if we are going to keep doing small single institution studies and then the criticisms are going to continue to be that we don’t know how to do LHR measurements, it makes you wonder if the LHR measurement is a valid measurement. If LHR can only be measured at 1 or 2 institutions, then perhaps we need to look at something else that can be standardized across institutions.
Liver position and lung-to-head ratio for prediction of extracorporeal membrane oxygenation and survival in isolated left congenital diaphragmatic hernia

Holly L. Hedrick, MD; Enrico Danzer, MD; Aziz Merchant, MD; Michael W. Bebbington, MD; Huaqing Zhao, MA; Alan W. Flake, MD; Mark P. Johnson, MD; Kenneth W. Liechty, MD; Lori J. Howell, RN, MS; R. Douglas Wilson, MD; N. Scott Adzick, MD
CONCLUSION: Liver position is the best prenatal predictor of outcome in isolated left congenital diaphragmatic hernia. Lung-to-head ratio alone should not be used to counsel families regarding mid gestational management choices.
Conclusions A variety of measures have been proposed as antenatal predictors of survival in CDH. Ultrasound parameters function at a similar level, whereas MRI determined parameters appear to offer better predictive value.
Conclusions: Evaluating total lung volumes is more accurate than is measuring only the contralateral lung size. Evaluating pulmonary vascularization (Cont-VI) is the most accurate predictor of neonatal outcome. Estimating the probability of survival and severe PAH allows classification of cases according to prognosis.
Michael Jordan 1988 Slam Dunk Contest on All-Star Weekend
PULMONARY HYPERTENSION

• Seems to be a big deal
• Sort of correlates with lung size
• May allow stratification of survival with severe disease
• Unclear the ideal way/place to measure
Russia again?????
MCGOON INDEX

• Assessment on risk of significant neonatal pulmonary hypertension
• Multiple studies have evaluated MRI/US in determining pulmonary blood flow and vessel size
• May only be relevant with severe disease
• With mild disease, though pulmonary hypertension is usually present, is usually not severe and life threatening
MCGOON INDEX

$$MGI = \frac{RPA_d + LPA_d}{Ao_d}$$

Normal $\geq 1.2$
3.7 + 2.8/4.8 = 1.35
4.2 + 3.3 / 5.4 = 1.39
Prenatal pulmonary hypertension index: novel prenatal predictor of severe postnatal pulmonary artery hypertension in antenatally diagnosed congenital diaphragmatic hernia.

Vuletin JF\textsuperscript{1}, Lim FY, Cnota J, Kline-Fath B, Salisbury S, Haberman B, Kingma P, Frischer J, Crombleholme T.

PURPOSE:
This study aim to assess the potential of prenatal predictors of postnatal severe pulmonary artery hypertension (PAH) in isolated left congenital diaphragmatic hernia (CDH) and to define a new prenatal pulmonary hypertension index (PPHI).
Prenatal pulmonary hypertension index: novel prenatal predictor of severe postnatal pulmonary artery hypertension in antenatally diagnosed congenital diaphragmatic hernia.


METHODS: A retrospective chart review of CDH patients between May 2005 and October 2008 was conducted. Ten patients with systemic-suprasystemic and 9 patients with subsystemic pulmonary hypertension at 3 weeks of age were identified. Diameters of the right pulmonary artery, left pulmonary artery (LPA(d)), aorta, and the length of vermis of the cerebellum were obtained from prenatal magnetic resonance imaging to calculate the PPHI \[=(\text{LPA(d)}/\text{length of vermis of the cerebellum}) \times 10\] and the modified McGoon index (MGI) \[=(\text{diameter of the right pulmonary artery} + \text{LPA(d)})/\text{diameter of aorta}\]. Prenatal pulmonary hypertension index and MGI were compared with lung-to-head ratio, percent predicted lung volume, and total lung volume for pulmonary hypertension and survival.
Prenatal pulmonary hypertension index: novel prenatal predictor of severe postnatal pulmonary artery hypertension in antenatally diagnosed congenital diaphragmatic hernia.


**RESULTS:**
The PPHI and MGI had a significant, negative correlation with pulmonary hypertension (r = -0.61, P = .005, and r = -0.72, P < .005, respectively). The PPHI and MGI are significantly lower in the systemic/suprasystemic PAH group compared with the subsystemic PAH group (1.11 +/- 0.32 versus 1.63 +/- 0.28, P = .004, and 0.71 +/- 0.15 versus 1.05 +/- 0.11, P < .001, respectively). There were no significant differences between the groups comparing the lung-to-head ratio, percent predicted lung volume, and total lung volume.

**CONCLUSION:**
Both PPHI and MGI accurately predict the severity of postnatal PAH in isolated left CDH.
Julius Erving 1977 classic dunk
PULMONARY HYPERTENSION

• Predicting the severity of this may not be helpful if the fetus is in a moderate to good prognosis category.

• May be more useful in determining outcomes in those with severe disease.

• If O/E LHR is < 26%
  • If dopplers were normal, survival was > 60%
  • If abnormal, survival was 0%

• Unclear what measurement is best.
$n = 41$
Overall survival rate 56.1%
Survivors: $n = 23$
Non-survivors: $n = 18$

$n = 31$
O/E-LHR

< 26%
Survival rate 45.2%
Survivors: $n = 14$
Non-survivors: $n = 17$

$n = 14$
PI

< 1.0 Z-s
Survival rate 71.4%
Survivors: $n = 10$
Non-survivors: $n = 4$

$n = 6$
PEDRF

< 3.5 Z-s
Survival rate 66.7%
Survivors: $n = 4$
Non-survivors: $n = 2$

$n = 17$

≥ 26%
Survival rate 90%
Survivors: $n = 9$
Non-survivors: $n = 1$

$n = 11$

> 3.5 Z-s
Survival rate 0%
Survivors: $n = 0$
Non-survivors: $n = 11$
SCORING SYSTEM
## Scoring system

<table>
<thead>
<tr>
<th></th>
<th>1</th>
<th>0</th>
<th>-1</th>
</tr>
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<tbody>
<tr>
<td><strong>Genetics</strong></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Karyotype</td>
<td>Normal (97%)</td>
<td>Mild (3%)</td>
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</tr>
<tr>
<td>Syndrome</td>
<td>No (98%)</td>
<td>Yes (2%)</td>
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<tr>
<td><strong>Cardiac</strong></td>
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<td></td>
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<tr>
<td>Congenital heart disease</td>
<td>None (89%)</td>
<td>VSD/ASD/Coarctation (9%)</td>
<td>Double outlet heart disease (2%)</td>
</tr>
<tr>
<td>LV/RV</td>
<td>No (97%)</td>
<td>Yes (3%)</td>
<td></td>
</tr>
<tr>
<td>McGoon</td>
<td>$\geq 1.2$ (33%)</td>
<td>$&lt; 1.2$ (67%)</td>
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<tr>
<td><strong>Hernia</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sac</td>
<td>Yes (5%)</td>
<td>No (95%)</td>
<td></td>
</tr>
<tr>
<td>Liver</td>
<td>Down (29%)</td>
<td>Up (71%)</td>
<td></td>
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<tr>
<td><strong>Lung</strong></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Lung-to-heart ratio</td>
<td>$\geq 1$ (66%)</td>
<td>$&lt; 1$ (34%)</td>
<td></td>
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<tr>
<td>PPLV</td>
<td>$\geq 15%$ (77%)</td>
<td>$&lt; 15%$ (23%)</td>
<td></td>
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<tr>
<td>Total lung volume</td>
<td>$\geq 18$ mL (73%)</td>
<td>$&lt; 18$ mL (27%)</td>
<td></td>
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</tbody>
</table>

The congenital diaphragmatic hernia congenital prognostic index scoring system consists of 4 groups: genetics, cardiac, hernia, and lung with 10 parameters. Each parameter and characteristics for scoring are depicted. The study population defined for each variable is represented by a percentage of available data.

LV/RV indicates left ventricle-to-right ventricle disproportion; VSD, ventricular septal defect; ASD, atrial septal defect; PPLV, percentage predicted lung volume.
SO, OTHER OPTIONS???

• Will discuss this in more detail later but
  • Postnatal surgery does pretty well
  • Isolated left sided CDH has approximately 70-90% survival
  • Usually with minimal long term morbidity
  • Less ECMO, waiting longer to do surgery....

• What about the “hidden mortality”
• What about in utero surgery??
Tommie Smith and John Carlos
1968 Summer Olympics
IN UTERO SURGERY

- Due to the poor outcome in high risk patients, attempts were made at in utero surgery for the highest risk group
- Open procedure with correction of the actual defect
- Complicated by early delivery
- High risk for the mother (general anesthesia, big uterine incision, multiple tocolytics...)
FETAL TRACHEAL OCCLUSION

- A significant amount of fluid will egress from fetal lungs during pregnancy
- A fetus with CHAOS will have over developed, over grown, over inflated lungs
- Theoretically, obstructing the flow out of the babies lung will result in expansion and potentially decreasing risk of pulmonary hypoplasia
- Can be done via minimally invasive technique
Balloon Tracheal Occlusion

Left lung expands by fluid buildup

Diaphragmatic hernia

Liver and small intestine pushed back down into abdomen
Michael Jordan buzzer beater in game 6 of NBA finals 1998
METHODS
Women carrying fetuses that were between 22 and 27 weeks of gestation and that had severe, left-sided congenital diaphragmatic hernia (liver herniation and a lung-to-head ratio below 1.4), with no other detectable anomalies, were randomly assigned to fetal endoscopic tracheal occlusion or standard care. The primary outcome was survival at the age of 90 days; the secondary outcomes were measures of maternal and neonatal morbidity.
CONCLUSIONS
Tracheal occlusion did not improve survival or morbidity rates in this cohort of fetuses with congenital diaphragmatic hernia.
Between Jan 2004 and Dec 2011, 150 fetuses with CDH identified

Classified at
- Severe: LHR < 1.0 and 1/3 of liver up
- Moderate: LHR > 1.0 and < 1.4 and 1/3 of liver up
- Mild: LHR > 1.4 with no liver

42 fetuses with severe CDH underwent FETO
- Survival = 54.8% with 47.6% having severe pulmonary arterial hypertension

In conventional, survival 5.9% in severe and 44.7% in moderate
- Severe pulmonary arterial hypertension in 90% of severe

Ruano R, et al Ultra OB/GYN, 2015;46(suppl. 1):31
Currently used criteria for TOTAL trial in left-sided CDH and the criteria used for right-sided CDH.

<table>
<thead>
<tr>
<th>Left-sided CDH</th>
<th>Right-sided CDH</th>
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<tbody>
<tr>
<td><strong>O/E LHR</strong></td>
<td><strong>Observational study</strong></td>
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<tr>
<td>Liver</td>
<td>Severe</td>
</tr>
<tr>
<td></td>
<td>&lt;25%</td>
</tr>
<tr>
<td>Liver “up”</td>
<td>Liver “up”</td>
</tr>
<tr>
<td><strong>RCT</strong></td>
<td><strong>RCT</strong></td>
</tr>
<tr>
<td>Total trial “severe”</td>
<td>Total trial “moderate”</td>
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<tr>
<td>Severe</td>
<td>Moderate</td>
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<tr>
<td></td>
<td>25—45.9%</td>
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<tr>
<td>Timing FETO:</td>
<td>If O/E LHR 25—34.9%: any position</td>
</tr>
<tr>
<td>between</td>
<td>If O/E LHR 35—44.9%: liver “up”</td>
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<tr>
<td>(weeks)</td>
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<tr>
<td></td>
<td>27&lt;sup&gt;±0&lt;/sup&gt; to 29&lt;sup&gt;±6&lt;/sup&gt; weeks</td>
</tr>
<tr>
<td></td>
<td>30&lt;sup&gt;±0&lt;/sup&gt; to 31&lt;sup&gt;±6&lt;/sup&gt; weeks</td>
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<tr>
<td></td>
<td>27&lt;sup&gt;±0&lt;/sup&gt; to 29&lt;sup&gt;±6&lt;/sup&gt; weeks</td>
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CDH, congenital diaphragmatic hernia; O/E LHR, observed/expected lung-to-head ratio; FETO, fetoscopic endoluminal tracheal occlusion. Ranges mentioned including the limits mentioned.
HERE’SSSSSSS
CINDY!!!

GIVE IT UP!!
CDH postnatal management

Cindy Gingalewski, MD
Objectives

▪ CDH prenatal predictors and postnatal management

▪ ECMO and CDH

▪ Current practice of minimally invasive surgical repair of CDH
Objectives

- CDH prenatal predictors and postnatal management
- ECMO and Cannulation techniques VV vs VA
- Current practice of minimally invasive surgical repair of CDH
CDH- prenatal predictors

Prenatal indicators that heighten awareness for ECMO

LHR: RIGHT lung area at the level of the 4 chamber view of heart/ head circumference

- LHR<0.6 = 100% mortality
- LHR>1.35 = 100% survival

O/E LHR takes into account lung growth later in gestation

- <25% = 18% survival, >45% = 89%

Liver up = 45%***

MRI fetal lung volume (34-35 EGA)

- Survivors 35 mL, ECMO 18mL, nonsurvivors 9mL
Ventilation strategy for CDH

Gentle ventilation strategy—utilize lowest pressure that provides adequate chest movement and gas exchange

<table>
<thead>
<tr>
<th>PARAMETER</th>
<th>GOAL</th>
<th>THRESHOLD</th>
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<tr>
<td>Preductal oxygen sats</td>
<td>&gt;90</td>
<td>&gt;85*</td>
</tr>
<tr>
<td>Postductal oxygen sats</td>
<td>&gt;70</td>
<td>&gt;60*</td>
</tr>
<tr>
<td>PaCO2</td>
<td>40-65</td>
<td>&lt;70</td>
</tr>
<tr>
<td>PIP</td>
<td>20-25</td>
<td>28</td>
</tr>
<tr>
<td>MAP</td>
<td>14-16</td>
<td>18</td>
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</table>

*Tolerate preductal sat > 85%, postductal >60% only if perfusion is satisfactory (e.g. non-acidotic and lactic acid not increasing).
Clinical Applications

- Cannot meet $O_2$ delivery requirements
- Inadequate end-organ perfusion
- Maximised inotropic/vasopressor support
- Failure of optimized ventilatory management
- Mortality $>80-90\%$ on current therapy
- Reversible underlying disease
ECMO

• Indications for ECMO:
  • Refractory hypoxemia

  • inability to maintain preductal sats > 85% (or using injurious settings to achieve satisfactory oxygenation, PIP > 28, MAP > 18)

  • Pressor resistant hypotension

  • Persistent metabolic acidosis

  • Inability to wean FiO2 from 100% in the first 48 hours
Objectives

▪ CDH prenatal predictors and postnatal management

▪ ECMO and CDH

▪ Current practice of minimally invasive surgical repair of CDH
What is ECMO?
The good oxygenation supplied by ECMO breaks the vicious cycle of PPHN, allowing the pulmonary hypertension to resolve and thus stopping the right to left shunting.
Contraindications

- Gestational age: <34 weeks
- Weight: <2kg
- Disease process: Must be REVERSIBLE
- Major bleeding complications: >Grade I IVH
- Serious or irreversible brain damage
- Lethal anomalies
Cannulation - Planning

- Target flow of 150 ml/kg/min or CI = 2.4

VV

VA

VV

Origen
VA ECMO for CDH

Pros
- Pulmonary and Cardiac Support
- Cannulate small infants
- Track record in CDH

Cons
- Carotid Ligation
- Cardiac stun
- Neurologic complications
VV ECMO for CDH

Pros
➢ No carotid ligation
➢ Deliver oxygenated blood to coronaries
➢ Preservation of pulsatile blood flow
➢ Less cardiac stun
➢ Less neurologic complications

Cons
➢ Lack of cardiac support
➢ Size limitations for DL cannula
The Data: VV vs. VA

➢ ELSO, 1991- 2006

➢ VA (2,257 / 86%), VV (371 / 14%)

➢ Survival: VA (50%), VV (54%) – P=0.03
   No difference after adjusting for severity

➢ VV-VA Conversion (138, 18%) – Survival 44% vs 54%
   in VV alone, P=0.001

➢ VA associated with more neurologic complications.
   VV associated with more renal complications

Long Term Outcomes

moderate to severe CDH: will go on to have long term morbidities falling into 3 categories:
• neurodevelopmental impairment in up to 25%
• chronic pulmonary hypertension or some degree of pulmonary injury (5%) leading to an increased incidence of asthma and recurrent pneumonias during the first 1-3 years of life
• feeding difficulties relating to oral aversion and/or gastroesophageal reflux.
Fetal therapy for CDH

- Began in 1980s
  - In utero repair for severe CDH
  - Outcomes- no improvement

- Tracheal plug trials
  - Show promise
  - Data not conclusive yet
Objectives

- CDH prenatal predictors and postnatal management
- ECMO and CDH
- Current practice of minimally invasive surgical repair of CDH
Minimally Invasive CDH Repair

CDHSG 1995-2010- 4516 CDH repaired.
151 MIS (3.4%). Only 20 centers.
26 laparoscopic, 125 thorascopic

Tsao et al J Ped Surg 46, 1158-1164, 2011
Laparoscopic Approach

▪ Better visualization of bowel

▪ Insufflation does not aid reduction

▪ Technically feasible but thoracoscopic preferred (0.6% vs 2.8% - CDHSG)
Thoracoscopic Approach

- Thoracoscopic
  - Reduction of hernia while visibility maintained
  - Gentle insufflation may lead to near-spontaneous reduction
Potential Limitations?

- Severe CDH
  - ECMO
  - Ventilatory requirements

- Size of defect
  - Need for patch
  - May need conversion to open

- CO2 insufflation and potential risk of acidosis
  - Great Ormond Street study - pCO2 96, pH 7.08
Predictors for open repair

- Stomach herniation
- Severe respiratory disease
  - Need for ECMO
  - Longer preoperative ventilatory days
What is the data?

- Retrospective reviews
  - Technically feasible
  - Longer operative times (30min-63min)
  - Conversion rate (5-25%)
    - Larger defects
    - Infant instability
  - Recurrence is higher after thoracoscopic repair (1-25%)
    - Not related to severity of disease or need for patch repair
  - Postoperative ventilator days and LOS shorter

- No difference in overall survival
  - CDHSG data with improved survival (82.9% vs 98.7%)

- Meta-analysis – similar conclusions (recurrence rate higher and OR time longer, similar survival)
Legacy/OHSU data

- 29 CDH repairs
- Recurrence 21% vs 7%
  - 1 patient had a second recurrence
    (thoracoscopic)
- Additional operative procedures 35 vs 43%
  - recurrence
  - gastrostomy tube
  - malrotation
  - incisional hernia
- Complications 55 vs 71%
  - no difference in bowel obstruction rates 6.9 vs 7.1%
Outcomes

- Overall survival 70-90%
  - 50% ECMO
- Respiratory difficulties
  - asthma- pulmonary htn
- Neurocognitive and language delays
- Oral aversion and GERD
  - Failure to thrive
Questions
SUMMARY

• CDH is still a major anomaly with significant morbidity and mortality
• Currently there is very limited ability to predict outcomes in a fetus, especially in a timely manner, to allow other pregnancy options such as termination
• Most predict morbidity but not mortality
• Probably, using multiple criteria will improve prediction
SUMMARY

• LHR is the most widely used ultrasonographic measure to stratify fetuses with CDH and values <1 associated with a higher risk of death, need for ECMO, and pulmonary hypertension at one month. **O/E is better than straight LHR**

• MRI parameters have better sensitivity and specificity for predicting survival at clinically used cut-off values than ultrasound, though are more costly. **WE SHOULD USE THIS MORE!!!!!**

• Echocardiographic measurements of the pulmonary vasculature, such as the modified McGoon index, have a high sensitivity and specificity for predicting death in fetuses with CDH.
SUMMARY

• Postnatal outcomes are continuing to improve
• Technology is allowing fetal surgery to be less invasive and therefore, with less maternal risks
• Preterm delivery is still a significant risk
• Currently in utero surgery is only indicated for severe disease but moderate may also be reasonable in the future
Ladybug Run
for CDH Awareness

2017

RACE DAY JULY 29TH
Lou Gehrig 1939 I am the luckiest man on earth speech after diagnosis of ALS
Jackie Robinson steals home in Game 1 of 1955 World Series
Roberto Clemente. Last game he played 1972
Don Larsen and Yogi Berra 1956 after Larsen pitched only perfect game in World Series History
Willie Mays World Series 1954
Juan Marichal 1965
Joe DiMaggio 1941. Nickname the Yankee Clipper
Dwight Clark  “The Catch” 1982
Joe Namath 1969 Pre-Super Bowl III
Usain Bolt 1988 Olympics after setting record in 200 meters