Congenital Diaphragmatic Hernia: Fetal Imaging Tips + Tricks

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CDH: Tips + Tricks

**IMPORTANT DECISIONS**, and tips to help make them

- Where is the hernia (and why do we care)?
- Is the liver up, and how much makes a difference?
- Is there a hernia sac?
- Is the patient a candidate for in utero therapy?
- Is there anything else I should look for?
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Hernia types

- Intrapleural
- Mediastinal
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Intrapleural hernias

90% of CDH cases
Incomplete closure of posterior pleuropertitoneal canal (Bochdalek)
Left 80-85%, right 10-15% and bilateral 2-5%
Complications: pulmonary hypoplasia, persistent PAH
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Mediastinal hernias: Ventral/Retrosternal
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Mediastinal hernias: Ventral, Morgagni-type
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Mediastinal hernias: Hiatal
TIP: stomach up without liver up on the right is a HH
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Why do we care if it’s intrapleural or mediastinal?

*Lung volumes make a difference*
Pulmonary Hypoplasia

How lung volumes are measured makes a difference

The Lung:Head ratio (LHR)

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Pulmonary Hypoplasia

How lung volumes are measured makes a difference

Predict mortality at >85%

LHR <1.0 (with liver up)

o/e-LHR <0.25 (L, liver up)

<0.45 (R)

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**Observed/Expected Total Lung Volume**

Use quality serial contiguous images.
Freehand ROI tool outlines higher-signal lung.
Sum areas from contiguous slices and multiply by slice thickness to obtain Total Fetal Lung Volume (TFLV).
Divide this observed value by the expected mean fetal lung volume for gestational age (= 0.0033 x GA^{2.86})^1 or fetal body volume^2.

O/E TFLV < 32% is poor

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US

stomach at level of 4-chamber view of heart
mediastinal shift
bowel in chest
scaphoid abdomen
liver in chest ventral to the stomach
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Herniated liver makes a difference

Liver herniation as predictor of outcome
Liver down: 79% survival
Liver up: 41% survival

In our series, %LH > 20 is predictive of increased mortality and the need for ECMO, independent of lung volumes.

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<table>
<thead>
<tr>
<th>Variable</th>
<th>cut-off value</th>
<th>Area under curve</th>
<th>95% confidence interval</th>
<th>Sensitivity / Specificity (%)</th>
<th>Accuracy (%)</th>
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</thead>
<tbody>
<tr>
<td>MORTALITY</td>
<td></td>
<td></td>
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<tr>
<td>LHR</td>
<td>&lt;1.40</td>
<td>0.70</td>
<td>0.57-0.84</td>
<td>65 / 71</td>
<td>69</td>
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<tr>
<td>o/e-LHR</td>
<td>&lt;0.41</td>
<td>0.72</td>
<td>0.59-0.85</td>
<td>68 / 69</td>
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<tr>
<td>o/e-TLV</td>
<td>&lt;0.32</td>
<td>0.78</td>
<td>0.67-0.89</td>
<td>78 / 74</td>
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<td>PPLV</td>
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<td>0.74</td>
<td>0.63-0.88</td>
<td>77 / 74</td>
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<tr>
<td>%LH</td>
<td>&gt;21%</td>
<td>0.75</td>
<td>0.62-0.85</td>
<td>72 / 79</td>
<td>77</td>
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<tr>
<td>LITR</td>
<td>&gt;14</td>
<td>0.72</td>
<td>0.55-0.86</td>
<td>67 / 81</td>
<td>77</td>
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<tr>
<td>LHR+%LH</td>
<td>0.75</td>
<td>0.75</td>
<td>0.61-0.89</td>
<td>73 / 72</td>
<td>75</td>
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<td>LHR+LITH</td>
<td>0.76</td>
<td>0.75</td>
<td>0.64-0.90</td>
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<td>76</td>
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<tr>
<td>o/e-TLV + LITR</td>
<td>0.80</td>
<td>0.80</td>
<td>0.70-0.90</td>
<td>79 / 81</td>
<td>80</td>
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<tr>
<td>PPV + %LH</td>
<td>0.80</td>
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A hernia sac makes a difference: 15% of cases

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The Treatment Options

Prenatal
- Termination
- No intervention
- Balloon occlusion of the trachea (FETO)

Delivery/Postnatal
- General supportive care
- ECMO, including EXIT-to-ECMO
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The Treatment Options

FETO
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The Treatment Options

FETO
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The Treatment Options

FETO: Fetal Endotracheal Occlusion

The preliminary data:

In O/E LHR <0.25, anticipate 15% (l up) – 30% (l down) survival if L-CDH  <5% with R-CDH

With FETO,  
52% survival for L-CDH  
29% for R-CDH

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The Treatment Options

FETO: Fetal Endotracheal Occlusion  TOTAL trial

- Isolated left intrapleural CDH for FETO b/t 27 and 30 weeks’ GA
- Severe cases
  - High risk of severe pulmonary hypoplasia \( o/e-LHR<0.25 \)
  - Liver herniation
  - High risk of pulmonary hypertension \( Cont-VI<20\% \)
- No other significant anomalies
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Imaging post intervention

Pre                                              post
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Gestational age at delivery makes a difference

An intervention is unlikely to help if it results in significant preterm delivery.

Technical advances have pushed delivery to late preterm. Elective balloon puncture or removal between 34-35 weeks.
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Associated abnormalities make a difference

<table>
<thead>
<tr>
<th>Affected System</th>
<th>Frequency</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiac</td>
<td>35%</td>
<td>Tetralogy of Fallot, VSD, coarctation, HLHS</td>
</tr>
<tr>
<td>Genitourinary</td>
<td>10%</td>
<td>Horseshoe kidney, crossed-fused ectopia, lower urinary tract obstruction</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>10%</td>
<td>Esophageal atresia/tracheoesophageal fistula, bowel atresia</td>
</tr>
<tr>
<td>Central nervous</td>
<td>7%</td>
<td>Holoprosencephaly, spina bifida</td>
</tr>
<tr>
<td>Pulmonary</td>
<td>&lt;5%</td>
<td>Sequestration, pleural effusion</td>
</tr>
<tr>
<td>Systemic ( Syndromes)</td>
<td>5-10%</td>
<td>Beckwith-Wiedemann, Fryns, Brachmann-de Lange</td>
</tr>
</tbody>
</table>
Clinical Features of Fryns Syndrome*

Broad nasal bridge or hypertelorism  
Hypoplastic lungs or abnormal lobation  
Diaphragmatic hernia, diaphragmatic anomalies  
Cardiac defects  
Coarse facial features  
Gastrointestinal tract anomalies, abnormal bowel fixation  
Distal digital hypoplasia  
Ear abnormalities  
Microretrognathia  
Macrostomia  
Genital anomalies  
Renal anomalies  
Polyhydramnios  
Central nervous system anomalies  
Cleft lip, cleft palate  
Talipes equinovarus  
Corneal abnormalities  
Hypoplastic nipples  
Cystic hygromas  
Osteochondrodysplasia  
Adrenal fusion†

*Listed in decreasing frequency of clinical presentation. This is a compilation of data from the reported cases in the literature.1–5,7–16
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Jugular vein size
