Prenatal Predictors of Postnatal CDH Outcome

Lisa K. Hornberger, MD
Fetal & Neonatal Cardiology Program
Department of Pediatrics
Department of Obstetrics & Gynecology
University of Alberta

Prenatally Diagnosed CDH

- Reported prenatal detection rates 20-100% over past decade
- Mean age at diagnosis 22-25 weeks
- Prenatal diagnosis more likely in left than right CDH
- Prenatal diagnosis more likely with advancing GA

Garne UOG 2002; 19: 329

Prenatally Diagnosed CDH

- Greater detection when multiple anomalies are present (50% vs 70%)
  - 55-65% isolated
  - 35-45% associated with additional abnormalities
    - 20-30% overall have multiple structural anomalies (CHD in 10-15%)
    - 10% chromosome abnormalities
    - Syndromes/Associations (e.g. Fryns, Cornelia de Lange, Meckel Gruber, Vater association)

Prenatal Diagnosis of CDH

- Goal of prenatal diagnosis & evaluation
  - more accurate counseling regarding morbidity & mortality
  - planned perinatal/neonatal care
  - development of prenatal intervention
Predictors of Outcome in Fetal CDH

- Worse prognosis in fetal CDH:
  - Associated anomalies
  - Right-sided CDH
  - Liver herniation (40-45% survival when liver up vs 75-93% when liver is down)
  - More severe lung hypoplasia

Fetal CDH: Predicting Pulmonary Hypoplasia

- Fetal ultrasound
  - General features
  - Lung area-head circumference ratio
  - 3D volumes
- Fetal echocardiography
  - Branch PA diameters
  - Pulmonary artery Doppler flow patterns
- Magnetic resonance imaging
  - Lung volume - total fetal body volume or other single biometric parameter

Fetal CDH: Predicting Pulmonary Hypoplasia

- Ultrasound-based LHR - Lung area to head circumference ratio (LHR)

Jani et al. UOG 2007; 30:67
Fetal CDH: Predicting Pulmonary Hypoplasia

- **U/S LHR - in left CDH**
  - Survivors
  - Nonsurvivors

Jani et al. UOG 2007; 30:67

Fetal CDH: Predicting Pulmonary Hypoplasia

- **U/S LHR - in left and right CDH**

Jani et al. UOG 2007; 30:67

Fetal CDH: Predicting Pulmonary Hypoplasia

- **U/S LHR - early vs late gestation**

Jani et al. UOG 2008; 31:37

Fetal CDH: Predicting Pulmonary Hypoplasia

- **LHR - early vs late gestation LHR**
  - False positive rate 10%
  - Sensitivity 44% at 22-23wks
  - 66% at 32-33wks

Jani et al. UOG 2008; 31:37
Fetal CDH: Predicting Pulmonary Hypoplasia

MRI for Determining Lung Volumes

Lung volume expected vs observed
- use of other biometric measurements
- use of calculated Total fetal body volume

Cannie et al Radiol 2006;241:847

Fetal CDH: Predicting Pulmonary Hypoplasia

MRI - fetal lung volume vs U/S LHR

Paek et al Radiol 2001;220:63

Fetal CDH: Predicting Pulmonary Hypoplasia

MRI - fetal lung volume to total fetal body volume

Cannie et al Radiol 2006;241:847

Fetal CDH: Predicting Pulmonary Hypoplasia

MRI - fetal lung volume/total fetal body volume (observed/expected) vs lung weight/body weight following pregnancy termination

Cannie et al Radiol 2006;241:847
Fetal Echocardiography in CDH

- Exclusion of congenital heart disease
- Assessment for risks of evolving hydrops (evidence of compression or altered ventricular filling)
- Assessment of lung mass & pulmonary vascular function
  - Branch PA diameters
  - Pulmonary arterial flow +/- maternal hyperoxia

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Sokol et al Am J OB/Gyn 2002;186:1085

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Branch PA diameters vs lung weight after pregnancy termination

Sokol et al Am J OB/Gyn 2002;186:1085
Fetal Echocardiography in CDH

Branch PA diameters correlate with morbidity


Duration on oxygen
Duration of ventilation
Length of stay

LPA (ga)
-0.69 0.009* -0.68 0.007* -0.77 0.002*

RPA (ga)
-0.43 0.139 -0.41 0.143 -0.41 0.160
Fetal Echocardiography in CDH

Branch PA diameters and serial assessment
- Of survivors:
  - All but 3 had normal LPA growth.
  - Of survivors with PA hypoplasia:
    - In 1: RPA grew normally; moderate postnatal morbidity
    - In 2: bilateral PA hypoplasia persisted; severe postnatal morbidity
      [DV (76, 108 days), DO (1 year each) & LOS (80, 154 days)]
- Of nonsurvivors:
  - All had ipsilateral PA hypoplasia ≤ 5th centile
  - 2 had bilateral branch PA hypoplasia

Sokol et al Am J OB/Gyn 2006;195:470

Fetal Echocardiography in CDH

Pulmonary arterial flow
- Spike and dome pattern during RV ejection
- Acceleration time to ejection time ratio (AT/ET) in some reports increases in others no change
  - LPA 0.17 ± 0.04
  - RPA 0.15 ± 0.04

Fuke et al Am J OB/Gyn 2003;188:228

Fetal Echocardiography in CDH

Pulmonary arterial flow

Lower AT/ET associated with increased PVR

Fuke et al Am J OB/Gyn 2003;188:228

Fetal Echocardiography in CDH

Intrapulmonary arterial blood flow

Contralateral

Ipsilateral

Moreno-Alvarez UOG 2008;31:164
Fetal Echocardiography in CDH

Intrapulmonary arterial blood flow

Moreno-Alvarez UOG 2008;31:164

Maternal hyperoxia challenge


- Assessed pregnancies at risk for fetal lung hypoplasia
- >30 weeks
- Assessment of changes in intrapulmonary blood flow - specifically pulsatility index
  Reactive if >20% change

14/29 total pregnancies nonreactive
11/14 neonatal demise
1/15 with reactive test died
Sensitivity 92%/specificity 82%
positive predictive value 79%
negative predictive value 93%
Fetal CDH

- Worse outcome associated with additional abnormalities, right CDH, liver herniation in several reports
- There is no one prenatal parameter which consistently predicts the degree of lung hypoplasia, and postnatal mortality and morbidity
- Need for prospective multicenter evaluation of predictors of outcome and generation of composite scores for best prediction