Doppler Assessment in Congenital Heart Disease and Fetal Heart Failure

Cristiano Jodicke, MD
Objectives

- Define fetal congestive heart failure
- Describe tools used to assess severity of CHF
- Discuss causes and treatment of fetal hydrops fetalis
Objectives

- Appropriate use of Doppler for fetal evaluation
- Demonstration of normal fetal Doppler patterns for umbilical artery, middle cerebral artery, ductus venosus and umbilical vein
- Abnormal fetal Doppler patterns – what do they look like and what does the information mean?
- Application in congenital heart disease
Congestive heart failure

Heart is unable to maintain sufficient blood flow to meet the needs of the body.

- Shortness of breath
- Exercise intolerance
- Body swelling
Fetal congestive heart failure

- Poor tissue perfusion
- Acidosis

Fetal death!!!
Factors affecting perinatal cardiac output

Parallel ventricular ejection

Stroke volume
- Preload
- Afterload
- Contractility
Fetal myocardium

- Less T-tubular system
- Less organized myofibrils in the fetus
- Differences in Ca uptake into the sarcoplasmic reticulum
- Decreased sympathetic innervation
  - smaller in size
  - mitochondria
  - sarcoplasmic reticulum
  - myofilaments,
  - alpha- and beta-adrenoceptors
  - t-tubuli

Fetal myocytes

- Myocardial growth -> increase in the number of muscle cells

Huhta et al. Seminars in Fetal & Neonatal Medicine (2005) 10, 542e552
Fetal myocardium

- Reduced ability of the fetal heart to contract and generate force
- Lower myocardial compliance
- Diminished Frank-Starling mechanism

Decreased cardiac reserve

Huhia et al. Seminars in Fetal & Neonatal Medicine (2005) 10, 542-552
Fetal myocardium

- Elevated venous pressure (reduce lymphatic flow)
- Elevated filling pressures
- Lower albumin concentration
  - plasma arginine vasopressin (decreases urinary production)
  - angiotensin II (increases fluid accumulation)
  - atrial natriuretic peptide (increases capillary permeability)
Hydrops!!!

- Edema with one or more effusions, or effusions in at least two spaces
  - Ascites
  - Pleural effusion
  - Pericardial effusion
  - Skin edema
Hydrops

Ascites
Pleural Effusion

Left

Pleural effusion

Right

Lung

Heart
Hydrops

Pericardial effusion
Hydrops

Skin edema
Differential Diagnosis
Cardiovascular

- **Malformation**
  - Left heart hypoplasia
  - Atrioventricular canal defect
  - Right heart hypoplasia
  - Closure of foramen ovale
  - Single ventricle
  - Transposition of the great vessels
  - Ventricular septal defect
  - Atrial septal defect
  - Tetralogy of Fallot
  - Ebstein anomaly
  - Premature closure of ductus
  - Truncus arteriosus

- **Tachyarrhythmia**
  - Atrial flutter
  - Paroxysmal atrial tachycardia
  - Wolff-Parkinson-White syndrome
  - Supraventricular tachycardia

- **Bradyarrhythmia**

- **High-output failure**
  - Neuroblastoma
  - Sacrococcygeal teratoma
  - Large fetal angioma
  - Placental chorioangioma
  - Umbilical cord hemangioma

- **Cardiac rhabdomyoma**
- **Other cardiac neoplasia**
- **Cardiomyopathy**
Differential Diagnosis

- **Chromosomal**
  - 45,X
  - Trisomy 21
  - Trisomy 18
  - Trisomy 13
  - 18q+
  - 13q-
  - 45,X/46,XX
  - Triploidy

- **Chondrodysplasias**
  - Thanatophoric dwarfism
  - Short rib polydactyly
  - Hypophosphatasia
  - Osteogenesis imperfecta
  - Achondrogenesis

- **Hematologic**
  - α-Thalassemia
  - Fetomaternal Transfusion
  - Parvovirus B19 Infection
  - In Utero Hemorrhage
  - Glucose-6-Phosphate Dehydrogenase (G6PD) Deficiency
  - Red Cell Enzyme Deficiencies

- **Thoracic**
  - Congenital Cystic Adenomatoid Malformation of Lung
  - Diaphragmatic Hernia
  - Intrathoracic Mass
  - Pulmonary Sequestration
  - Chylothorax
  - Airway Obstruction
  - Pulmonary Lymphangiectasia
  - Pulmonary Neoplasia
  - Bronchogenic Cyst
Differential Diagnosis

➢ **Twin Pregnancy**
  - Twin-twin transfusion syndrome
  - Acardiac twin

➢ **Infections**
  - Cytomegalovirus
  - Toxoplasmosis
  - Parvovirus B19 (Fifth Disease)
  - Syphilis
  - Herpes
  - Rubella

➢ **Malformation Sequences**
  - Noonan Syndrome
  - Arthrogryposis
  - Multiple Pterygia
  - Neu-Laxova Syndrome
  - Pena-Shokeir Syndrome
  - Myotonic Dystrophy
  - Saldino-Noonan Syndrome

➢ **Metabolic**
  - Gaucher Disease
  - GM1 Gangliosidosis
  - Sialidosis
  - Mucopolysaccharidosis Type 4a

➢ **Urinary**
  - Urethral Stenosis or Atresia
  - Posterior Urethral Valves
  - Congenital Nephrosis (Finnish)
  - Prune Belly Syndrome

➢ **Gastrointestinal**
  - Midgut Volvulus
  - Malrotation of the Intestines
  - Duplication of the Intestinal Tract
  - Meconium Peritonitis
  - Hepatic Fibrosis
  - Cholestasis
  - Biliary Atresia
  - Hepatic Vascular Malformations
Diagnostic Approach

- Maternal history
- Ultrasonography
- Fetal echocardiography

Labs:
- T&S
- screen for hemoglobinopathies
- Kleihauer-Betke
- TORCH titers

- Fetal karyotype
- Screening for lysosomal storage diseases
Causes of fetal congestive heart failure

- Fetal arrhythmias
- Anemia
- Congenital heart disease with valvular regurgitation
- Non-cardiac malformations such as diaphragmatic hernia or cystic hygroma
- TTTS
- Atrioventricular fistula with high cardiac output
Team Approach

• Perinatologists
• Cardiologists
• Neonatologists
Fetal heart assessment

- Contractility
- Cardiac size
- Arterial/ Venous Doppler
- Four-valve Doppler
- Myocardial performance
Doppler
Arterial Doppler Waveform Analysis
Hemodynamic Basis

Maulik, Arbeille, Kadado. Biol Neonate, 1992
Umbilical Artery Doppler

End Diastolic Flow

Doppler Analysis – the Indices

- Resistance Index (RI)
- Pulsatility Index (PI)
- Systolic/Diastolic Ratio (S/D)
- Diastolic Average Ratio (D/A)

Maulik et al, 1984

RI = (S–D)/S (Pourcelot, 1974)
PI = (S–D)/A (Gosling, 1978)
S/D Ratio (Stuart and Drumm, 1980)
D/A Ratio (Maulik et al, 1982)
# Umbilical Artery Doppler Absent/Reverse End-Diastolic Flow

Compiled from 1126 reported cases

<table>
<thead>
<tr>
<th>Perinatal Outcome</th>
<th>Mean</th>
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<tbody>
<tr>
<td>Perinatal mortality</td>
<td>45%</td>
</tr>
<tr>
<td>Gestational age</td>
<td>31.6 wks</td>
</tr>
<tr>
<td>Cesarean for fetal distress</td>
<td>73%</td>
</tr>
<tr>
<td>Apgar score at 5min &lt;7</td>
<td>26%</td>
</tr>
<tr>
<td>Admission to NICU</td>
<td>84%</td>
</tr>
<tr>
<td>Congenital anomalies</td>
<td>10%</td>
</tr>
<tr>
<td>Aneuploidy</td>
<td>6.4%</td>
</tr>
</tbody>
</table>

Umbilical Artery Doppler in CHD

- Obstetric complications, e.g. fetal growth restriction
- Increased abnormality in CHD with:
  - Aneuploidy
  - Other malformations
- Isolated CHD
  - Most cases: no specific change
  - Higher UA pulsatility index in right heart obstructive lesions, e.g. pulmonary stenosis, pulmonary atresia, TOF, severe Ebstein’s, tricuspid atresia

Copel et al, JUM, 1999; Ursem et al, UOG 2001; Donofrio et al, 2003; Kaltman et al, UOG, 2005
Middle Cerebral Artery Doppler

Arbeille et al, UMB, 1987
Fetal Cerebral Artery Doppler in CHD

- MCA Doppler in hypoplastic left heart
  - Decreased CV flow impedance
  - Lower MCA Doppler indices
- MCA Doppler in right sided obstructions
  - >> left sided obstructive lesions
  - > normal
- Cerebral Doppler/umbilical Doppler (CPR, U/C)
  - Controversial - ?“brain sparing effect” in CHD
  - Higher (Donofrio 2003) vs. normal (Rychik 2005)
- ? Predictor of neurological outcome in CHD
Absent or reverse A wave

- Right heart dysfunction
- Late predictor fetal decompensation
- Independent predictor of mortality in FGR
- Independent predictor of intact survival (neonatal)


Kiserud: Dop US Ob Gyn, (Ed:Maulik), Springer. 2005
Fetal Ductus Venosus Doppler in CHD

- Absent or reverse “A” wave indicates fetal cardiac failure and predicts the outcome including mortality
- Absent or reverse “A” in 1st Trimester predicts 30-40% of euploid CHDs
- Increased peak velocities in severe outflow tract obstructions (HLHS, PS, AS, TOF, Ebstein’s, TV atresia)
- Abnormal DV in CHD with other malformations, aneuploidy, fetal growth restriction
- No significant changes in most cases of CHD
Advanced fetal decompensation
Reflects right heart dysfunction with increased right atrial pressure
Poor prognosis
Pulsed Doppler of Mitral/ Tricuspid Valve

Low E/A ratio

- Impaired ventricular diastolic function
  - Decreased preload
  - Decreased ability active myocardial relaxation
  - Valvular dysfunction
Figure 16  E/A ratios as a function of gestational age. Source: ref. 33.
Tricuspid regurgitation
Mitral regurgitation
Outflow tracts

Pulmonary

Aortic
Cardiac output

\[ \text{Stroke Volume} \times \text{FHR} \]
Stroke Volume = V x A

- Volume = Velocity time integral (VTI)
- Area = Cross-sectional area of the valve

Area = $\pi r^2$
Normal Heart Size
Cardiomegaly

Universal sign of heart failure

C/T area ratio = (normal 0.2 - 0.35).
C/T circumference ratio = normal < 0.5
Measurements of ventricular wall thickness

Any left ventricular posterior wall thickness greater than or equal to 4 mm is abnormal.
Myocardial performance index

Systolic and Diastolic function

- independent of heart rate
- independent of ventricular geometry
- non invasive

Index = \( \frac{a - b}{b} = \frac{(ICT + IRT)}{ET} \)

Ventricular dysfunction

- Prolongation of Isovolumetric contraction time (ICT)
- Prolongation of Isovolumetric relaxation time (RCT)
- Reduction of ejection time (ET)

ICT + IRT

ET

Tei index
How to calculate the Tei index

\[
\frac{(a - b)}{b}
\]

Normal

Left < 0.76
Right < 0.65

Ichizuka et al, Early Hum Develop 2005;81:273-279
Fractional shortening is constant throughout gestation (normal > 0.28)

- Myocardial compromise
- Increase in the fetal ventricular workload
**Fetal cardiovascular profile score**

<table>
<thead>
<tr>
<th></th>
<th>Normal</th>
<th>-1 point</th>
<th>-2 points</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Hydrops</strong></td>
<td>None (2 pts)</td>
<td>Ascites or Pleural effusion or Pericardial effusion</td>
<td>Skin edema</td>
</tr>
<tr>
<td><strong>Venous Doppler</strong></td>
<td>UV</td>
<td>UV</td>
<td>UV pulsations</td>
</tr>
<tr>
<td>(Umbilical vein)</td>
<td>DV (2 pts)</td>
<td>DV</td>
<td></td>
</tr>
<tr>
<td>(Ductus venosus)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Heart Size</strong></td>
<td>&gt;0.20 and &lt;0.35 (2 pts)</td>
<td>0.35 - 0.50</td>
<td>&gt; 0.50 or &lt;0.20</td>
</tr>
<tr>
<td>(Heart Area / Chest Area)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Cardiac Function</strong></td>
<td>Normal TV &amp; MV</td>
<td>Holosystolic TR or RV/LV S.F. &lt; 0.28</td>
<td>Holosystolic MR or TR dP/dt &lt; 400 or Monophasic filling</td>
</tr>
<tr>
<td></td>
<td>RV/LV S.F. &gt; 0.28</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Biphasic diastolic filling (2 pts)</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Arterial Doppler</strong></td>
<td>UA (2 pts)</td>
<td>UA (AEDV)</td>
<td>UA (REDV)</td>
</tr>
<tr>
<td>(Umbilical artery)</td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>

CVP score of ≥8 is associated with a good perinatal outcome

A cardiovascular profile score in the surveillance of fetal hydrops

Hofstaetter C, Hansmann M, Eik-Nes SH, Huhta JC, Luther SL

Human fetal cardiovascular profile score and neonatal outcome in intrauterine growth restriction
Mäkikallio K, Räsänen J, Mäkikallio T, Vuolteenaho O, Huhta JC.

Cardiovascular profile score in those who died was 5
Management

- Prognosis
- Gestational age
- Presenting signs and symptoms
- Fetal therapy???
Medical treatment of fetal heart failure

- Prolonging pregnancy
- Preventing prematurity and prenatal asphyxia
Management

- Consultation with neonatology
- Consultation with cardiology
Management

- Myocardial dysfunction
- Arrhythmias
- High output states (fistula and anaemia)
- Cardiomegaly
- Decreased ventricular shortening

Heart failure will often occur outside 60 and 200 beats/min.

Digoxin

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Management

- Anti-Rho and anti-La antibodies
  - Valvular regurgitation
  - Heart block
  - Myocardial dysfunction
  - Effusion

Dexamethasone 4 mg daily

Huhta et al. Seminars in Fetal & Neonatal Medicine (2005) 10, 542-552
CREASY AND RESNIK’S MATERNAL-FETAL MEDICINE: PRINCIPLES AND PRACTICE, SIXTH EDITION
Management

- TTTS
- Acardiac twins

Laser treatment

Huhta et al. Seminars in Fetal & Neonatal Medicine (2005) 10, 542-552
Management

- Anemia

Transfusion

Huhta et al. Seminars in Fetal & Neonatal Medicine (2005) 10, 542-552
Management

- Aortic stenosis
- Hypoplastic Left Heart Syndrome and Intact or Highly Restrictive Atrial Septum

In utero balloon angioplasty

Biophysical Profile
Management

Delivery time???
Conclusions

- Heart failure can be diagnosed in fetuses and the severity can be estimated.

- Serial studies with emphasis on venous Doppler can be useful in management.

- Transplacental treatment of fetal heart failure could result from accurate diagnosis and fetal/maternal stratification.
Conclusions

• Fetal Doppler findings in CHD depend on:
  • Associated malformations
  • Aneuploidy
  • Obstetrical complications, e.g. fetal growth restriction, preeclampsia

• Fetal arterial Doppler has selective utility in CHD.

• Fetal venous Doppler has defined utility in CHD:
  • Absent or reverse “A” wave in the ductus venosus indicates fetal heart failure and poor prognosis.
  • Umbilical venous pulsations indicate poor prognosis.
Thank You