Fetal Congestive Heart Failure: Diagnosis, Management, and Prognosis

James C. Huhta, M.D.
Perinatal Cardiology
Thursday Nov. 3, 2016 2:15 PM

Faculty Disclosure Information
In the past 12 months, I have no relevant financial relationships with the manufacturer(s) of any commercial product(s) and/or provider(s) of commercial services discussed in this CME activity.

I do not intend to discuss an unapproved/investigative use of a commercial product/device in my presentation.

Fetal Congestive Heart Failure

Fetal Supraventricular Tachycardia

CV Profile
10-point score

Hydrops
None (2 pts)
Anuria or Pleural effusion or Pericardial effusion

Venous Doppler
Normal (2 pts)
UVDV (Umbilical vein)
DV pulsations

Heart size
Normal (2 pts)
< 0.35
0.35 - 0.50
> 0.50 or < 0.20

Cardiac function
Normal TV & MV
AVV 0.5 - 0.20
Monophasic filling
Biphasic diastolic filling
Holosystolic TR
RV/LV S.F. > 0.28
TR dP/dt < 400

Arterial Doppler
Normal (2 pts)
UA (Umbilical artery)
(2 pts)
UA (AEDV)
UA (REDV)

Fetal congestive heart failure
CV profile Score

1. Hydrops
2. Venous Doppler
3. Heart size
4. Heart function (SF, AVV regurg, etc.)
5. Arterial Doppler

Detection of CHD-Disproportion
Fetal Congestive Heart Failure
Abnormal Venous Doppler


Perinatal Management
Salvage of HLHS

Fetal Valve Regurgitation

Tricuspid regurgitation dP/dt

Perinatal Management
Cardiomyopathy

Myocarditis
Genetic syndromes
Inherited defects
Consider transplantation as a neonate

Fetal CHF with CHD
Examples

CHD with increasing heart size in utero
Tet absent valve syndrome
Pulmonary atresia with collaterals
Ebstein's malformation
Critical AS
L isomerism with CHB
**Fetal CVP Score - 146 fetuses**

**Congenital Heart Disease Perinatal Mortality**

<table>
<thead>
<tr>
<th>Score Group</th>
<th>Sensitivity</th>
<th>Specificity</th>
<th>PPV</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-2</td>
<td>0.25</td>
<td>0.98</td>
<td>0.88</td>
</tr>
<tr>
<td>3-5</td>
<td>0.22</td>
<td>0.98</td>
<td>0.75</td>
</tr>
</tbody>
</table>

**Sensitivity** | **Specificity** | **PPV**

**For Mortality**

- Sensitivity: 0.25
- Specificity: 0.98
- PPV: 0.88

**For 5 minute Apgar score <=6**

- Sensitivity: 0.22
- Specificity: 0.98
- PPV: 0.75

---

**Diagnosis of Fetal CHF in IUGR**

**IUGR – longitudinal observations (≤ 32 weeks)**

- Umbilical artery
- Ductus venosus
- Short term variation
- Middle cerebral a.

**Validation of CVP score IUGR-Makikallio et al.**

- Eight out of 75 neonates died before discharge or had severe CP (n=2)
- Delivery at earlier gestational age 28 (range 24-35) weeks vs. 35 (range 26-40) weeks, p<0.001
- Lower fetal CVP scores 4 (range 2-6) vs. 9 (range 5-10), p<0.001
- All fetal subset scores of CVP except umbilical artery evaluation were lower (p<0.001) in the group with neonatal death.
Validation of CVP score-IUGR

- Neonates with 5-minute Apgar scores < 7 had lower CVP scores than with scores > 7 (6 (2-10) vs. 9 (5-16), p<0.001).
- Umbilical artery NT-proANP levels of newborns with CVP score < 6 were greater (5208 (2850-16030) pmol/L) than the levels of neonates with CVP exceeding 6 (1626 (402-9574) pmol/L), p=0.0001.
- All NT-proANP values of newborns with CVP score < 6 were above the 95th percentile NT-proANP value in normal pregnancies, while 42 out of 67 (63%) fetuses with CVP > 6 showed NT-proANP concentrations exceeding the 95th percentile value in normal pregnancies.
- Umbilical artery NT-proANP values correlated inversely and significantly with CVP score values.

Validation of CVP score-Complete AV Block

- We have implemented a strategy that includes the biophysical profile, which assesses fetal well-being, in combination with the cardiovascular profile that assesses cardiac function and the circulation.
- Two cases of fetal complete heart block in which early delivery was recommended due to worsening cardiovascular profile scores. Biophysical profile scores were normal. Both babies were successfully treated, despite having risk factors that predicted poor outcomes. We hypothesize that our management protocol initiated intervention before fetal compromise, hydrops, and myocardial damage occurred.
- We recommend an evaluation of heart function in addition to an assessment of fetal well-being in fetuses with complete heart block. Early delivery should be considered if there is evidence of distress and/or deteriorating cardiac function.

Validation of CVP score-T-T Transfusion

- Assess the relationship between cardiomyopathy and recipient twin (RT) outcome in twin-twin transfusion syndrome (TTTS).
- Fetal echocardiography and outcomes data in 62 consecutive pregnancies with TTTS were reviewed. The primary outcome was neonatal RT survival.
- The severity of RT cardiomyopathy at presentation was assessed by the cardiovascular profile score (CVPS). RT outcomes and odds of survival were compared between groups stratified by CVPS.
- Overall neonatal survival for all fetuses was 61% (76 of 124). RT survival was 58% (36 of 62). Grouped by CVPS, RT survival was greater (50%) for those with CVPS >/= 9 and even higher (74%) for CVPS of 10.
- Atrioventricular valve regurgitation was associated with negative RT outcome. Other factors at presentation were not predictive of RT Outcome.
- A normal CVPS in the RT in TTTS is predictive of improved survival compared with an abnormal CVPS, even in RTs with minor deductions. Standard clinical staging did not predict outcome.

Standard clinical staging did not predict outcome.

**Validation of CVP score**
The role of echocardiography in prenatal diagnosis of pulmonary arteriovenous malformation

CVP score 7/10
Effusion, cardiomegaly (area ratio 0.47), decreased SF 0.27

Sinkovskaya E, Berkley E, Bogdan D, Sclater A, Abuhamad A. The role of echocardiography in prenatal diagnosis of pulmonary arteriovenous malformation
Prenatal Diagnosis. 2009; 29:634-636

**Validation of CVP score - Sacrococcygeal Teratoma**
To determine the value of CVP Score for the prognosis of the fetus with SCT

26 fetuses with SCT;
CVP score of survivors was 9 (range 8-10)
CVP in nonsurvivors 7 (range 4-9) p<0.004

Respondek et al. ISUOG Abstract Sept. 2005

**Validation of CVP score - Cardiomyopathy**

• Toronto data per Dr. Jaeggi
• Fetal cardiomyopathy Multiple causes with a similar phenotype Hypertrophic CM with higher risk
• Diastolic dysfunction is the key finding
  – Monophasic inflow, increased IRT, venous decreased early diastolic flow, A reversal in DV
• CVP Score is useful
  – Increased mortality with CVP score LT or equal to 7 (RR 5.4 95% CI 1.5-20)
  – Increased mortality with hydrops (RR 2.3m 95% CI 1.4-3.9)
Perinatal Management
Cardiomyopathy

Digoxin Therapy for the fetus in CHF

Structural cardiac defects (n= 21) or noncardiac anomalies (n=7)
Length of treatment (5.0 ± 3.2 weeks)


Results-Digoxin treatment group

• Mortality was 32%, and limited to those with moderate (3/13) and severe (6/9) CHF.
• A CVPS of ≥ 6 was the best predictor of survival (sensitivity 0.83, specificity 0.75).
• All fetuses that died in utero had notching of the umbilical venous flow.
• The first, last and CVPS after 1 week of treatment predicted survival (Odds ratio 2.34, 95% CI 1.10-4.96).

Results of digoxin Rx

• The overall CVPS increased from baseline during treatment (p = 0.003) in all subjects.
• The CVPS score is useful in assessing therapeutic effects of digoxin in the fetus with multiple etiologies for CHF

Summary: CVPS Associated with Mortality

<table>
<thead>
<tr>
<th>Group</th>
<th>Perinatal Mortality</th>
<th>CVP Score</th>
<th>Mortality* or Early Delivery**</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 (CHD)</td>
<td>20%</td>
<td>&lt;7</td>
<td>*87.5% vs. 15.2%</td>
</tr>
<tr>
<td>2 (Hydrops)</td>
<td>60%</td>
<td>&lt;7</td>
<td>*73.5% vs. 26.5%</td>
</tr>
<tr>
<td>3 (IUGR)</td>
<td>11%</td>
<td>&lt;6</td>
<td>100%</td>
</tr>
<tr>
<td>4 (AVB)</td>
<td>*82%, **26%</td>
<td>&lt;7</td>
<td>*100%, **100%</td>
</tr>
<tr>
<td>5 (Digoxin)</td>
<td>32%</td>
<td>&lt;5</td>
<td>100%</td>
</tr>
</tbody>
</table>
Future Research

- Disease – specific CVP Score
- Comparison with Biophysical Profile Score
- First Trimester CVP Score
- Mouse embryo CVP Score