Delivery Planning for the Fetus with Congenital Heart Disease

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Objectives

♥ Review the current outcome literature that shows the benefit of prenatal diagnosis of congenital heart disease
♥ Learn which congenital heart defects may lead to in-utero compromise
♥ Learn which congenital heart defects may lead to immediate postnatal compromise
♥ Discuss the appropriate postnatal intervention strategies available to improve the outcome of babies born with specific congenital heart defects
Fetal Cardiology: The Diagnostic Dilemma

♥ Role of the fetal cardiologist is to identify the fetus with congenital heart disease and improve outcome, both prenatally and after delivery

♥ Outcome, in part, depends on the success of transition from in-utero to postnatal life, and depends on both the anatomy and predictions of postnatal physiology
  • Cardiac diagnosis
  • Ventricular function
  • Arrhythmias
  • Pulmonary Vascular Resistance
  • Patency/size of the DA
  • Patency/size of the FO

257 with CHD/49 diagnosed prenatally in Berlin

Prenatal diagnosis care plan:
  • Initiation of appropriate management in the DR and immediate transfer to CICU after delivery

Postnatal diagnosis:
  • 27% had been already discharged at time of diagnosis
    • Acidosis and cardiac dysfunction: 50%
    • Murmur or other exam findings: 50%

Fuchs, et al

Fetal Diagnosis CHD: Improved Outcome

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Fetal Diagnosis CHD: Improved Outcome

♥ Prenatal vs. Postnatal Diagnosis:
  • Higher preop O2
  • Fewer cases with cardiac failure
  • Fewer cases with preop ductus closure
  • Shorter duration of postop ventilation
  • Shorter stay in the CICU
  • Less post-discharge heart failure

*Improved Survival*

Fuchs, et al

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Fetal Diagnosis D-TGA: Improved Outcome

♥ 318 with TGA/ 68 diagnosed prenatally in France

♥ Prenatal vs. Postnatal Diagnosis
  • Quicker admission to ICU
  • Shorter hospital stay
  • Less hemodynamic distress (acidosis, organ failure)
  • Lower preop mortality despite similar management
    • 0 vs. 6%
  • Lower postop mortality despite similar surgical risk
    • 0 vs. 9%

*Improved Survival*

Bonnet, et al

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Fetal Diagnosis HLHS: Improved Outcome

Tworetzky, et al

♥ 88 patients with HLHS/ 33 diagnosed prenatally
♥ Prenatal vs. Postnatal Diagnosis:

♥ Preop factors correlating with mortality
• Postnatal diagnosis
• Acidosis, need for bicarb or inotropes
• Ventricular dysfunction
• Survival: 100% vs. 66% (pre vs. postnatal dx)

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Fetal Diagnosis CHD: Postnatal Management

Mirlesse, et al

♥ 229 fetuses with CHD in France
• 110 births; 60 transferred at delivery/30 with compromise
• TGA: 74% required immediate transport
  • 3/23 required balloon septostomy in the delivery room
• Rhythm abnormalities: 53% required immediate transport
  • 5/7 with CHB and 4/6 with atrial flutter required pacing

♥ Proposed Management Guidelines:
• Babies with TGA or significant arrhythmias should be delivered in a specialized unit where immediate intervention can be performed

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### CHD: Lesion Specific Considerations

**Ductal Dependent Pulmonary Blood Flow:**

**Hypoplastic Right Heart Syndrome**

<table>
<thead>
<tr>
<th>Cardiac lesion</th>
<th>Special considerations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Left-to-right shunt lesions (ventricular septal defect, atrioventricular canal defect)</td>
<td>None</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td></td>
</tr>
<tr>
<td>Transposition of the great arteries</td>
<td></td>
</tr>
<tr>
<td>Total anomalous pulmonary venous return</td>
<td></td>
</tr>
<tr>
<td>Single ventricle with ductal-dependent pulmonary blood flow</td>
<td></td>
</tr>
<tr>
<td>Single ventricle with ductal-dependent systemic blood flow</td>
<td></td>
</tr>
</tbody>
</table>

None, may require PGE if severe right ventricular outflow tract obstruction is present.

Lower than normal saturations expected. Deliver in a center that can do balloon atrial septostomy (BAS) if atrial septal restriction suspected. PGE₂ in most cases.

Lower than normal saturations expected. Deliver in a center that can perform urgent cardiac surgery if obstruction expected.

Lower than normal saturations expected. PGE₂ to be started. Deliver in a center that can perform urgent cardiac surgery or BAS if atrial septum restrictive.
### Ductal Dependent Systemic Blood Flow: Hypoplastic Left Heart Syndrome

![Ultrasound images of a baby's heart](Image)

**CHD: Lesion Specific Interventions**

**Johnson, et al.**

<table>
<thead>
<tr>
<th>Cardiac lesion</th>
<th>Anatomy/Physiology</th>
<th>Intervention</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypoplastic left heart syndrome with intact atrial septum</td>
<td>Inability to provide pulmonary venous return (oxygenated blood) to systemic circulation</td>
<td>Surgical atrial septectomy or balloon atrial septostomy</td>
</tr>
<tr>
<td>Transposition of the great arteries with restrictive atrial septal defect</td>
<td>Inability to provide pulmonary venous return (oxygenated blood) to systemic circulation</td>
<td>Balloon atrial septostomy</td>
</tr>
<tr>
<td>Total anomalous pulmonary venous return with obstruction</td>
<td>Inability to provide pulmonary venous return (oxygenated blood) to systemic circulation</td>
<td>Surgical repair</td>
</tr>
<tr>
<td>Tetralogy of Fallot with absent pulmonary valve</td>
<td>Severely dilated main and branch pulmonary arteries causing external bronchial compression</td>
<td>Prone positioning, continuous positive airway pressure, or intubation</td>
</tr>
<tr>
<td>Complete congenital heart block</td>
<td>Poor cardiac output due to lack of atrioventricular synchrony or low ventricular rate</td>
<td>Increase ventricular rate with isoproterenol or pacing</td>
</tr>
</tbody>
</table>
Babies with CHB are at increased risk for mortality with most deaths occurring in-utero or in the neonatal period. The cardiac evaluation of these fetuses is difficult. Protocol:
- Biophysical profile to assess fetal well-being
  - Breathing
  - Movement
  - Tone
  - Amniotic fluid volume
  - Heart Rate Reactivity
- Cardiovascular profile to assess cardiac function
Heart Block: Fetal Assessment

Donofrio, et al

Fetal CHB: Case #1

Donofrio, et al

- **24 weeks:** CHB at 50bpm
- **31 weeks**
  - Enlarged heart (-1 CVP)
- **33 weeks**
  - Enlarged heart (-1 CVP)
  - New TR (-1 CVP)
- **35 weeks**
  - Enlarged Heart (-1 CVP)
  - Decreased LV function, TR, new MR (-2 CVP)
  - Decreased fetal movement
Fetal CHB: Case #2

- 28 5/7 weeks: CHB at 40bpm
  - Small pericardial effusion (-1 CVP)
  - UV pulsations (-1 CVP)
  - Enlarged heart (-1 CVP)
  - Mild TR (-1 CVP)

- Maternal terbutaline given

- 29 weeks
  - No change in effusion, new skin edema (-2 CVP)
  - UV pulsations (-1 CVP)
  - Enlarged heart (-1 CVP)
  - Decreased LV function, worsening TR (-2 CVP)

Heart Block: Fetal Assessment

- Cardiac dysfunction may be an early predictor of in-utero compromise in fetuses with CHB
- The evaluation of fetuses with CHB should include serial assessment, including evaluation of both fetal wellness and cardiac function
- Delivery planning should be based on the complete fetal assessment which includes degree of fetal compromise and cardiac dysfunction

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The Foramen Ovale: Impact in Fetuses with CHD

Donofrio, et al

370 fetuses; 105 with CHD
- Group I- obligate right to left atrial shunt (HRHS)
- Group II- obligate left to right atrial shunt (HLHS)
- Group III- CHD at risk for LA hypertension (TGA, AS)

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Number of fetuses</th>
<th>Number with FO R/C</th>
<th>Number with hydrops</th>
<th>Number of deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total: Fetals</td>
<td>370</td>
<td>6 (2%)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Total: Normals</td>
<td>224</td>
<td>0</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Group I: HRHS</td>
<td>15</td>
<td>2 (13%)</td>
<td>2 100%</td>
<td>1 50%</td>
</tr>
<tr>
<td>Group II: HLHS</td>
<td>20</td>
<td>2 (10%)</td>
<td>0 0%</td>
<td>1 50%</td>
</tr>
<tr>
<td>Group III: AS/TGA</td>
<td>9</td>
<td>2 (22%)</td>
<td>0 0%</td>
<td>2 100%</td>
</tr>
<tr>
<td>Total: Groups I-III</td>
<td>44</td>
<td>6 (14%)</td>
<td>2</td>
<td>4 67%</td>
</tr>
</tbody>
</table>

The risk of foramen ovale restriction/closure is greater in fetuses with CHD

Foramen ovale restriction/closure in fetuses with CHD can result in distress in-utero or at birth

Fetuses with HRHS and obligate right to left atrial shunting are more likely to develop hydrops fetalis

Serial evaluation should be performed for at risk fetuses

These fetuses may be candidates for intervention
Hypoplastic Left Heart Syndrome

- 0.21/1000 live births; 1% of infants with CHD
  - Associated restrictive/closed ASD 6-11%

HLHS: Restrictive/Intact ASD
- 38 patients; 26 diagnosed prenatally
- Fetal course
  - 8 had progression of disease with increased restriction from first to last fetal study
- Overall early survival: 68%

Glatz, et al
HLHS: Restrictive ASD

- 41 fetuses with HLHS/Restrictive ASD
- Pulmonary vein Doppler
  - S, D, A waves
- 20% required emergent atrial septoplasty
  - Fetal PV Doppler forward/reverse flow predictor of emergent atrial septoplasty
  - Sensitivity = 0.88; Specificity = 0.97

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HLHS: Postnatal Intervention

Heterotaxy: Complex Single Ventricle

Interrupted IVC,
Azygous continuation of the IVC to the SVC...
Heterotaxy: Complex Single Ventricle

- Levocardia, rightward stomach
- Double outlet RV with ventricular inversion and small RV
- Unbalanced AV canal to the LV with common atrium and large VSD
- Aortic atresia with hypoplastic aortic arch
- Normal SVC with interrupted IVC and azygous continuation to SVC
- Intact atrial septum
- Normal pulmonary venous drainage into LA
- Obstructed pulmonary venous return with a tiny LA decompressing vein

LA decompressing vein

LA hypertension, Obstructed pulmonary vein flow

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Heterotaxy: Complex Single Ventricle

- HLHS variant
- Intact atrial septum
- Obstructed pulmonary venous drainage
- Normal SVC with interrupted IVC and azygous continuation of the IVC to the SVC

Heterotaxy with Intact Atrial Septum: Postnatal Intervention

- Planned delivery by C/S with rapid catheter intervention to open the atrial septum
- Details of anatomy important for intervention planning (HLHS/IAS and interrupted IVC)
  - Input from Delivery team, ICU team, Cath team
- Planned prophylactic ECMO to stabilize circulation for the procedure
  - Deliver first with ECMO ready vs. EXIT to ECMO
  - Risk vs. Benefit Analysis
    - Risk to mother vs. benefit to fetus

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D-TGA

♥ 0.24/1000 live births; 5-7% of infants with CHD

Normal Circulation  Transposition

TGA: Foramen Ovale and Ductus Arteriosus

Foramen Ovale  Ductus Arteriosus

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Fetal TGA Circulation

• Increased O2 content of blood in the PAs and DA
  • Decreased PVR
  • Ductal restriction

Why don’t all fetuses with TGA get in trouble?

• Anatomic differences of the foramen ovale
• Different response of the pulmonary arteries and ductus to increased oxygen
• Umbilical vein flow
  • Percent of highly saturated UV flow that crosses the DV in the fetus is variable (ranging between 20 and 80%)
  • The more highly oxygenated the blood that crosses the foramen ovale and reaches the ductus and PAs, the more ductal constriction and PA dilation
D-TGA: Management

- Prostaglandin
- Rashkind Balloon Septostomy

TGA: Impact of DA and FO

- 130 patients with a prenatal diagnosis of TGA
- FO/DA described using criteria of Maeno
  - Foramen ovale
    - Abnormal if septum primum bulged > 50% to the LA
    - Abnormal if the angle of septum primum < 30%
    - Abnormal if there was lack of swinging motion of septum primum
  - Ductus arteriosus
    - Abnormal if small with narrowest diameter z score < -2SD
    - Abnormal flow
      - Continuous aorta to PA
      - Bidirectional

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Fetal findings:
• Abnormal FO in 19%
• Abnormal DA in 5%

Outcome
• Overall Mortality of 3%
  • 2 shortly after birth despite aggressive resuscitation
  • 2 post-operative

Fetal Predictors of Outcome
• Critical condition at birth
  • 29% with a restrictive FO and/or DA vs. only 6% with a normal FO/DA
• Restrictive FO and DA in 4 patients
  • All required intervention with 2 deaths
    – Case 1: cyanosis/severe acidosis; died at 3 days with brain injury
    – Case 2: failed initial balloon septostomy; successful at 4 hours; died of brain injury
• Specificity for critical condition
  • Abnormal FO or DA: 84%
  • Abnormal FO and DA: 100%
TGA Fetus: A Case

**Prenatal course:**
- 20 weeks: “Normal 4-chambered heart”
- 34 weeks:
  - TGA, no FO, no DA
  - Fetal distress
  - Emergent C/S in OR adjacent to cardiac OR

**Postnatal course:**
- Presentation: Limp, poorly perfused, No HR
- Intervention
  - Immediate ECMO via open chest
  - Arterial switch on day 7

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TGA Case: Intact FO/No DA

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Summary

♥ Fetal diagnoses of CHD improves outcome by identifying defects that may lead to compromise and allowing time for adequate planning of services for in-utero or postnatal intervention

♥ Defects requiring neonatology intervention
  • Ductal dependent lesions

♥ Defects requiring immediate cardiac intervention
  • TGA with restrictive ASD
  • HLHS with restrictive ASD
  • Obstructed TAPVR
  • Unstable arrhythmias: some tachycardias, CHB

Summary

♥ Improved outcomes are dependent on a team approach for management
  • OB/MFM
  • Fetal Cardiology
  • Neonatology
  • Postnatal Cardiology Team:
    Cardiac Intensivists, Cardiologists, EP Specialists, Interventional Cath, CV Surgery
Children’s National Heart Institute