Fetal Cardiac Interventions: Pulmonary Atresia/IVS and Aortic Stenosis

Italian Society For Pediatric Cardiology
October 16th, 2013
Padova, Italy

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Introduction

• Pulmonary Atresia with IVS
  – Hypoplastic Right Heart Syndrome

• Aortic Stenosis / HLHS in the fetus

• Research Presentation
Fetus with Pulmonary Atresia with IVS “Hypoplastic Right Heart Syndrome”
PAIVS 24w
TR  Ductus Retrograde Flow
20w TV Dysplasia + PA (Ebstein’s like)
PAIVS – Hypoplastic Right Heart Syndrome

- Three broad categories

- 1. “Best” Good size RV, TV and PV

  - Achieve biventricular repair with standard postnatal therapy

  - Perforation and balloon of PV

  - +/- BTS / RVOT muscle **resection**

  - **Probably do not need fetal intervention**
PAIVS

• 2. “Worst” Very Small RV, TV and infundibular atresia
  
  – Coronary stenosis or atresia

  – “RV dependent” coronaries

  – Not candidates for fetal intervention
PAIVS

3. Borderline right heart

- Definitely need BTS and RVOT muscle resection
- Wait for right heart structures to grow
- May achieve a 2V repair
- May need a 1 ½ V repair
- May need a Fontan
Pulmonary Atresia with IVS
Outcomes After Fetal Diagnosis

Table 1: Fetal PA/IVS: Demographics and Post-Natal Outcomes, 1990-2004

- Fetal diagnosis PA/IVS: n=36
  - Live birth: n=25
    - Study group: n=23
    - Fetal intervention: n=2
    - Termination: n=10
    - Fetal demise: n=1
      - Biventricular repair: n=7
      - Fontan: n=9
      - Glenn: n=5
      - Patent RV outflow tract: n=3
      - Closed RV outflow tract: n=2 (RV/DCC)
      - BTS: n=2
      - Death: n=2

Salvin J Pediatrics
PAIVS
Fetal Predictors of Postnatal 2V Repair

Figure 1: TV z-score at early and late fetal echo as indicator of biventricular repair

TV z-score at early and late fetal echo
Achievement of biventricular repair
PAIVS Fetal Intervention
Technical Challenges

• More complex RV geometry

• Smaller ventricle (target)

• RVOT is behind the sternum

• * Free to perforate the PV (no coronaries)
• * Free to perforate the RVOT and PAs
• * Coronary flow not as compromised
22w fetus with Pulmonary Atresia with IVS
Percutaneous Fetal Intervention

Fetal PAIVS Needle in RVOT

Fetal PAIVS Wire Across
Pulmonary Atresia with IVS
In Utero Balloon Dilation at 22w

Fetal PAIVS Balloon Inflation

Fetal PAIVS Post Color Flow
PAIVS
Needle to RV  Cannula at PV
PAIVS
Balloon      Color Doppler
In Utero Valvuloplasty for Pulmonary Atresia With Hypoplastic Right Ventricle: Techniques and Outcomes
<table>
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<th>Result</th>
<th>Outcome</th>
<th>Current</th>
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<td>26</td>
<td>-</td>
<td>liveborn 1V</td>
<td>s/p BDG</td>
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<tr>
<td>2</td>
<td>26</td>
<td>-</td>
<td>liveborn 1V</td>
<td>s/p BDG</td>
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<tr>
<td>3</td>
<td>23</td>
<td>-</td>
<td>TOP</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>23</td>
<td>+</td>
<td>alive</td>
<td>s/p BDG</td>
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<td>22</td>
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<td>biventricular</td>
</tr>
<tr>
<td>6</td>
<td>24</td>
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<tr>
<td>7</td>
<td>27</td>
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</tr>
<tr>
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<td>+++</td>
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<tr>
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</tr>
<tr>
<td>14</td>
<td>26</td>
<td>++</td>
<td>liveborn Balloon only</td>
<td>biventricular</td>
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Fetal Aortic Stenosis/HLHS
Fetal Aortic Stenosis at 20 w – “mild”
Fetal Aortic Stenosis – 20 w
Monophasic MV Inflow Doppler
Fetal Aortic Stenosis 20w
Aortic arch antegrade flow
Fetal Aortic Valve Stenosis and the Evolution of Hypoplastic Left Heart Syndrome
Patient Selection for Fetal Intervention

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“Mild” Aortic Stenosis

- 2 weeks later
Aortic Stenosis – Pre Balloon
Severe LV Dysfunction
Aortic Stenosis – Pre Balloon
Aortic arch retrograde flow
Fetal Aortic Stenosis - Pre Balloon
Aortic Stenosis Jet
Fetal Aortic Valvuloplasty
Percutaneous Technique

- Ultrasound guided
- Percutaneous
- Epidural anesthesia
- Fetal positioning
- Fetal anesthesia
- 19G needle
- 0.014” wire
- Coronary balloon
AMNIOTIC FLUID

MATERNAL ABDOMINAL WALL

UTERINE WALL

FETAL CHEST & RIBS

RV

LU

MV

LA

AORTIC VALVE
LINE UP

1) ENTRY SITE ON ABDOMEN
2) LV APEX
3) LV OUTFLOW TRACT
SLOWLY ADVANCE NEEDLE TO LV APEX
ADVANCE NEEDLE INTO LU CAVITY
REPOSITION NEEDLE TO LUOT
REMOVE STYLET
MAY GET BLOOD RETURN
0.014" Floppy Wire
Make sure it is in the aorta
Position balloon across the annulus.

Coronary balloon

NinJa 2.75mm

Raptor 2.75mm

The balloon should be pre-loaded on the wire.
RETRACT CANNULA
SLIGHTLY TO GIVE
ROOM FOR BALLOON
INFLATION
DEFLATE + RETRACT BALLOON INTO CANNULA + REMOVE
FETAL BRADYCARDIA
EARLY
1) IM EPINEPHRINE OR ATROPINE
2) INTRACARDIAC RV OR LV.
EPINEPHRINE OR ATROPINE
22G NEEDLE
HEMOPERICARDIUM
DRAIN WITH
EITHER 19G OR 22G

* OBSERVE FOR 15 MIN
AFTER PROCEDURE FOR
BRADYCARDIA + HEMOPERICARDIUM
26w Aortic Valvuloplasty - Percutaneous U/S Guided Needle Course
23w Fetal Aortic Valvuloplasty - Percutaneous
23w Aortic Valvuloplasty
Wire and Balloon Placement

Wire in aorta - Balloon inflated
Fetal AS – 1 day post balloon AOV
AOV flow
Fetal Aortic Stenosis – 5 weeks Post Balloon of AOV
Improved LV function
Fetal AS, post balloon of AOV
Late gestation
Postnatal

- Repeat balloon dilation of the AOV
- Biventricular circulation
- Now 6 years old and doing well
In contrast

24 weeks AS  36 weeks HLHS

LV / RV     LV / RV
S/p Fetal Aortic Valvuloplasty – Echo at 8 Years

Repeat aortic valvuloplasty at 18 months
Gradient reduced from 70 to 35 mm Hg
Tech Successful and live born then ~ 50% are biventricular
In-utero Balloon for Fetal AS – HLHS: Results

• Postnatal mortality

• Biventricular from birth 0/29

• HLHS 11/46 (24%)

• 1V – 2V rehab/conversion 2/7
In-utero Balloon for Fetal AS – HLHS: Results: Recent Data 2008-2011

- Reflects learning curve
- Improved patient selection
- Technical success 24/28 (86%)
- Fetal demise 2/28 (7%)
- Biventricular outcome 17/26 (65%)
The Future of FCI for AS

• To correctly identify patients

• Destined to evolve into HLHS

• High likelihood of benefiting from FCI and achieve and good biventricular repair

• Avoid FCI in those that are too far advanced and will not benefit
Fetal Resuscitation

Bradycardia CPR
Fetal Resuscitation

Epinephrine to RV

FHR recovery
s/p Fetal Aortic Valvuloplasty
Aortic Regurgitation
Resolution of Aortic Regurgitation after Fetal Balloon Aortic Valvuloplasty

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Introduction

- In neonates and children post-dilation AR often progresses
- In our early experience with fetal intervention, we observed that post-dilation AR is relatively common
- The outcome of post-dilation AR in fetuses is unknown
- We studied the fate of AR in fetuses undergoing BAV for AS
Methods - Patients

Attempted BAV in utero (n=90)

Technically unsuccessful BAV (n=21)

Technically successful BAV (n=69)
Fetuses
n=69

No or Mild AR
n=46

Moderate or Severe AR
n=23

Technically Successful BAV
AR Grade post-BAV
Results - Variables associated with AR

- Lower GA at the time of the procedure  \( p = 0.002 \)
- Higher balloon-annulus ratio  \( p = 0.002 \)
- Aortic annulus Z-score  \( p = 0.79 \)
- Ascending aorta Z-score  \( p = 0.31 \)
Variables associated with AR:

GA at the time of BAV

p = 0.002
Variables associated with AR:
Balloon-Annulus Ratio

*Balloon-Annulus Ratio*
Technically successful BAV (n=69)

Fetal demise (n=6)
Still in utero (n=3)

Live-born (n=60)
Aortic Regurgitation
Change during gestation
(60 live-born infants)

Post-BAV

Moderate or severe AR (n=18)
Mild AR (n=20)
No AR (n=22)

At birth

Moderate or severe AR (n=2)
Mild AR (n=8)
No AR (n=49)
Resolution of Aortic regurgitation at birth

Moderate AR post BAV

Absence of AR at birth
Resolution of Aortic Regurgitation

- Mild AR
- Moderate or Severe AR

Days after the procedure vs. AR persistence (%)
Discussion – Fetal AR

1. Why does fetal AR appear to be well tolerated?
   1. Low systemic resistance – placenta
   2. High LVEDP in fetus with AS – mitigates the degree of AR
   3. RV is providing circulatory support

2. Why does AR improve post-BAV?
   1. Unique hemodynamics result in less mechanical stress on the valve
   2. Fetal valves are dynamic, contain proliferating cells with an immature phenotype
   3. Underlying pathologic stenosing process
Thank You!

Grazie!