Predicting Biventricular Repair in the Fetal Borderline Left Heart

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20 weeks

“Mild” aortic stenosis
2 weeks later at 22 weeks
Severe LV dysfunction
5 weeks post fetal balloon dilation of the aortic valve
Fetal AS/HLHS

23w  36w
Fetus   ➔   Neonate

• Fetus
  – Progressive disease (usually worse, not better)
  – Time critical decisions
  – Imperfect published data
  – No magic formula
  – Fetal therapy an option for a small subgroup with AS

• Neonate
  – Depends how you look at it!
  – Institutional variation or management
  – Option of a staged LH growth strategy
Which one has HLHS?
23 weeks
Which one has HLHS? 23 weeks
Left Heart Adequacy

• Not just about the size of LH structures

• Systolic and diastolic function

• Endo and myocardial fibrosis

• Valve function
  – Particularly in the borderline LH
Fetus – Neonate – Child – Adult

• Children are not small adults

• Fetus is not a child

• Fetal myocardium
  – Exquisitely sensitive to loading conditions
  – Responds differently to insults
  – Instead of “heart failure”
  – Redirection of flow / Irreversible damage / Growth arrest

– Potential for recovery?
Fetus: Left Heart Hypoplasia

4 groups of LHH

- Aortic +/- mitral stenosis
  - Short, globular shaped, EFE

- Coarctation/Shone’s complex
  - Long and narrow LV

- Complex CHD: Unbalanced AVC, heterotaxy

- Underfilled LV
  - Ebstein anomaly
  - Congenital diaphragmatic hernia
Fetal AS with evolving HLHS

Arch Retrograde Flow
Two common types of Left Heart Hypoplasia
If you are a hypoplastic LV
Better to be “long and narrow” vs “short and globular”
Ventricles can expand
Circumferentially more than longitudinally
Left Heart “Growth” Paradox
Fetus - Neonate

• Uniqueness of the fetal circulation

• Due to neonatal changes

• Small in the fetus can become normal after birth

• Dilated in the fetus can become small after birth
Familiar Scenario: Diagnosis of HLHS “Develops a Life of Its Own”

- Fetal diagnosis of small left heart structures
- PGE started at birth
- LH structures look small
- Dx of HLHS is perpetuated
Familiar Scenario: Diagnosis of HLHS
“Develops a Life of Its Own”

• Fetal echo
  – Apex forming skinny LV
  – Biphasic MV inflow
  – Antegrade flow around the arch
  – No EFE
  – Possibly small PFO (reduced R-L flow)
  – LSVC to CS

• Neonate
  – Increased PBF with filling of LH
  – Time and patience
  – Possibly a COA repair
Left Heart Hypoplasia with a “long and skinny” LV
20 weeks GA – Hypoplastic LV?
20 weeks GA– Hypoplastic Aortic Arch
20 weeks GA–Hypoplastic Left Heart Structures
20 weeks GA – Aorta to Pulmonary Size Discrepancy
36 w – Hypoplastic Left Heart Structures
COA/Shone’s – 1 week of age
Left CDH – Left Heart Hypoplasia?
Why is the LH Hypoplastic in CDH?

- External compression?
- Decreased trans-foramenal flow?
- Decreased pulmonary venous return (pulmonary hypoplasia)
CDH – 3 months of age
Why does the LHH improve postnatally?

• Lung expansion
• Improved pulmonary blood flow
• Increased preload
• No inherent MV, myocardial or AOV disease
• Hibernating LH structures
Significance and outcome of left heart hypoplasia in fetal congenital diaphragmatic hernia

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The Pendulum Keeps Swinging
Two Questions
In a fetus with Aortic Stenosis and a Dilated, Dysfunctional LV

• 1. Will this fetus develop HLHS at birth?
  – There is data

• 2. If we perform a technically successful fetal aortic valvuloplasty....will this fetus go on to have a biventricular circulation after birth?
  – Developing data
The Spectrum of Fetal Aortic Stenosis

- Severe
- Mild

- Neonatal CAS "Normal"
- Hypoplasia
- Severe HLHS

Stenosis

20w 30w 40w
Fetal AS with evolving HLHS

Arch Retrograde Flow
The natural history of the hypoplastic left heart syndrome.

Allan LD, Sharland G, Tynan MJ.

Department of Perinatal, Guy’s Hospital, London, U.K.

In a fetus, examined initially at 22 weeks gestation, we identified the echocardiographic features of a dilated, hypertrophied and poorly contracting left ventricle. The presumptive diagnosis was critical aortic stenosis. Subsequent scans at 32 weeks and at term showed that the left ventricle had not grown since the first study such that the left ventricle had developed the appearance of a hypoplastic and densely echogenic chamber. Thus, in some forms of the hypoplastic left heart syndrome, the left ventricle can be of normal size or even dilated in early pregnancy. This may mean that the more subtle sign of poor left ventricular contraction could be overlooked in a routine four-chamber view obstetric scan.

Fig. 1. Left ventricular long-axis dimension.

McCaffrey and Sherman: Fetal Aortic Stenosis

Fetal Aortic Stenosis → HLHS
Simpson and Sharland 1997

Figure 2  Graph of left ventricular end diastolic volume versus gestational age in fetuses with aortic stenosis. 1 Balloon dilatation of aortic valve performed in utero following first scan. 2 Failed attempt at balloon dilatation of aortic valve in utero (case 9).
Fetal Aortic Valve Stenosis and the Evolution of Hypoplastic Left Heart Syndrome
Patient Selection for Fetal Intervention

Kaarin Mäkikallio, MD, PhD; Doff B. McElhinney, MD; Jami C. Levine, MD; Gerald R. Marx, MD;
Steven D. Colan, MD; Audrey C. Marshall, MD; James E. Lock, MD;
Edward N. Marcus, MSc; Wayne Tworetzky, MD
Fetal Aortic Valvuloplasty Percutaneous Technique

- Ultrasound guided
- Percutaneous
- Epidural anesthesia
- Fetal positioning
- Fetal anesthesia
- 19G needle
- 0.014” wire
- Coronary balloon
↑ Normal LV  ↑ Dilated LV with AS
Technique #1 – Percutaneous
20w Fetal Aortic Stenosis

AS jet 20 weeks ~ 1mm jet width  Fetal Anesthesia
23w Aortic Valvuloplasty
U/S Guided Needle Course
6 dimensional procedure

- 3 spatial dimensions
- A beating heart
- A moving fetus – floating in fluid
- A target that moves when you hit it

And then add the human element...several of them
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 Aortic Valvuloplasty Transuterine 20w

Needle to LV

Wire Across

Balloon Inflation
Fetal AS and EFE 23 week GA
Acute Change in AoV Flow
23w - Post Aortic Valvuloplasty

AS jet 23 weeks
Pre valvuloplasty
~ 1mm jet width

AS jet 23 weeks
Post valvuloplasty
~ 2.5mm jet width
S/p Fetal Aortic Valvuloplasty – Echo at 8 Years

Repeat aortic valvuloplasty at 18 months
Gradient reduced from 70 to 35 mm Hg
Fetal Intervention for Aortic Stenosis
Pre balloon 23 w        Post balloon 26w
Fetal Int for AS
Echo at 18 months. Post AOV repair
Fetal Aortic Valvuloplasty: Does it work?

- Depends on how you look at the data
World Cup 2010
USA vs. England
Attempted fetal aortic valvuloplasty n=93
Technically Successful n=72/93

- Tech Success and Live Born n=64
  - Comfort care, n=1
    - Died from sepsis, n=1
    - Died post-transplant, n=1
  - Rx HLHS from birth n=38
    - Converted to 2V after BDG N=5
      - ACHIEVED BIVENTRICULAR CIRCULATION n=31

- Fetal Demise n=6
  - In Utero n=2

- Biventricular from birth n=26

Median age at follow-up: 4 yrs (0.1 - 11)
Median age 2.3 yrs (all after BDG)
Update April 2012
s/p Fetal Aortic Valvuloplasty
Aortic Regurgitation
Fetal Intervention for AS
Aortic Regurgitation

- Attempted procedures n=92
- Technically successful 71/92 (77%)
- Aortic regurgitation
  - None 1/3, mild 1/3, mod-severe 1/3
- Fetuses with mod-severe were younger
- Larger BAR associated with more severe AR (>1.2)
- AR had no association with fetal demise
- 85% were born with none or mild AR

- 48% Technically successful and live born have achieved a biventricular repair
Limitations/Failures of Fetal Interventions

1) Technical Limitations
   • Fetal positioning
   • Fetal stabilization / movement
   • Imaging
   • Equipment inadequacies

2) Biological Limitations
   • No animal model
   • Late referral
   • Patient selection
   • Non uniform postnatal approach
“Borderline Left Heart” ?
Thank you!

- World Congress of
- Pediatric Cardiology and Cardiac Surgery
- Cape Town South Africa Feb 2013
Reasons Not to Perform Fetal Interventions
Not enough data either way
Back to the Drawing Board

• Put all fetal cardiac interventions on hold

• Perform a multicenter natural history study

• Create animal models for defect and technique

• Perform a prospective randomized study

• Should it become clinical care?
Ventricular Hypoplasia

• Clearly adequate and clearly inadequate

• How do we decide what needs a decision?
  – When it invokes a reaction “Is this 1V or 2V?”

• Why the obsession?
  – Univentricular circulation has a poor long term outcome

• We need to better understand the CHDs we are treating
Left Heart Hypoplasia
Physiologic and/or Anatomic

• Anatomic
  – Cavity size
  – Valves size
  – Myocardium (myocardial fibrosis and EFE)

• Physiologic
  – Cavity size: might depend on loading conditions
  – Myocardium: systolic and diastolic function
  – Valves: stenosis and/or regurgitation
Fetus: Left Heart Hypoplasia
Why is it so important?

• May be progression from mild to severe LHH

• Appropriately counsel the parents about the likely outcome and prognosis

• May choose TOP

• Delivery planning

• Potential for fetal therapy
Neonate: Left Heart Hypoplasia
Why is it so important?

- We are used to instant gratification in pediatric cardiology

- Do we really need to make the “big decision” at birth?

- We need a more thoughtful, innovative and patient approach
2V vs. 1V

• 1970s-1980s
  – HLHS palliation not available or poor survival
  – Patients pushed to 2V – high mortality

• 1990s-2000
  – Improving results for HLHS palliation
  – Maybe doing a Norwood on everyone
2 Strategies to add to standard Rx

• Fetal intervention

• Standard Rx (still struggle with 1V vs. 2V)

• Left heart rehabilitation surgery

• Maybe we will look at some patients differently

• There are many ways to look at the same patients or data!