Non-Invasive Imaging Modalities in Single Ventricle Hearts

(Modalités d'imagerie non invasive simples coeurs ventricule)

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Edmonton, CANADA
Disclosure

- Consultant role with Novartis Inc. (modest)
- Je suis tout à fait incapable de communiquer en français

pardon!

ma faute
Overview

• Why?
  - Surgical/Interventional Decisions

• How?
  - Which modalities most useful in 2015?
  - Which are still in development?
Can Imaging Make a Difference?

Plan Surgical Decisions: Will my patient survive a Single Ventricle Palliation?

Intention to Treat: Preoperative

Interstage (includes surgical mortality)

“Intermediate Term” (includes early post-Fontan surgical mortality)
Mechanisms of Single Ventricle (RV) dysfunction?

Which technique for which problem?

- **Myocardial Function**
  - RV maladaptation to systemic pressures
  - Effects from neonatal bypass ischemia
  - Inadequate coronary reserve, chronic ischemia
  - Dyssynchronous contraction

- **AV Valve function**

- **Circulatory/Extracardiac issues**

- **Anatomic Repair (Shunt, Aortic Arch)**

- **Pulmonary Veins**

- **Echo/MRI**

- **3D-Echo**

- **MRI/Other**

- **Cath**

- **ALL**

Hauser et al. Pediatr Cardiol 2003
Friedberg et al. JASE 2007
Tan et al. Am Heart J 2007
Kriakides et al. Int J Cardiol 2007
I. Rebeyka  D. Ross

- Current Norwood (Sano) STS surgical mortality is < 5% for HLHS
- BTS Era mortality, (pre-2003) was much higher

Atallah et al, Circulation, 2008
7 month old, HLHS, birthweight 2.1 kg.
Norwood Sano in 2nd Week of life
Poor weight gain, persistent cyanosis.
Persistently diminished RV function
Being considered for Stage II.
Has never left the hospital
**ECHO:** Moderate RV dysfunction: FAC 20%
Will the patient survive to a Glenn?

“echocardiographic findings were not independent risk factors for interstage mortality”

(J Thorac Cardiovasc Surg 2012;144:896-906)
## Will the patient survive to a Glenn?

<table>
<thead>
<tr>
<th>Echo Modality</th>
<th>Interstage Survival OR</th>
<th>p-value</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>RV EDV (ml/m²)</td>
<td>1.01</td>
<td>0.13</td>
<td>Difficult to measure</td>
</tr>
<tr>
<td>RV ESV (ml/m²)</td>
<td>1.02</td>
<td>0.03</td>
<td>Marginal significance</td>
</tr>
<tr>
<td>RVEF (%)</td>
<td>1.0</td>
<td>0.01</td>
<td>47 ± 8 vs 42 ± 7</td>
</tr>
<tr>
<td>RV FAC (%)</td>
<td>0.97</td>
<td>0.12</td>
<td>Hard to explain why</td>
</tr>
<tr>
<td>Pulmonary vein flow reversal present</td>
<td>0.46</td>
<td>0.18</td>
<td>Not a quantitative measure</td>
</tr>
<tr>
<td>AV valve regurgitation grade</td>
<td>1.87</td>
<td>0.06</td>
<td>More predictive if moderate or severe</td>
</tr>
</tbody>
</table>
Will the patient survive to a Fontan?

Impact of Initial Shunt Type on Cardiac Size and Function in Children With Single Right Ventricle Anomalies Before the Fontan Procedure
The Single Ventricle Reconstruction Extension Trial

<table>
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<tr>
<th>RV systolic size and function</th>
<th>Transplant-Free Survivors</th>
<th>Transplanted or Died</th>
<th>Hazard Ratio</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>End-systolic volume, ml/BSA^{1.3}</td>
<td>221</td>
<td>49.88 ± 16.74</td>
<td>12</td>
<td>99.61 ± 53.29</td>
</tr>
<tr>
<td>End-systolic area, cm²/BSA^{0.8}</td>
<td>284</td>
<td>15.82 ± 4.07</td>
<td>17</td>
<td>20.98 ± 8.09</td>
</tr>
<tr>
<td>Ejection fraction, %</td>
<td>221</td>
<td>43 ± 7</td>
<td>12</td>
<td>34 ± 9</td>
</tr>
<tr>
<td>Area fraction, %</td>
<td>284</td>
<td>33 ± 7</td>
<td>17</td>
<td>26 ± 8</td>
</tr>
<tr>
<td>Peak systolic velocity, cm/s</td>
<td>291</td>
<td>5.33 ± 1.44</td>
<td>17</td>
<td>5.81 ± 1.23</td>
</tr>
<tr>
<td>Isovolumic contraction acceleration, cm/s²</td>
<td>156</td>
<td>35.24 ± 11.57</td>
<td>8</td>
<td>32.50 ± 8.80</td>
</tr>
</tbody>
</table>

A larger, poorer functioning RV at 14 months of age is associated with Death or Transplant:

J Am Coll Cardiol 2014;64:2026–35
Customizing the Assessment of the Single Ventricle

Jeffrey Smallhorn

Nee Khoo          Tim Colen          Edythe Tham
Can we customize the assessment of the RV?

Assessing Contractile Function

- **Ventricular myocardium fibre architecture:**
  - RV has 2 “layers” of myofibers instead of 3.
  - Lack mid “layer” of circumferential fibers.
  - Unsuitable to act as a systemic ventricle.

Epicardial  Endocardial
Background

• Systemic RV lacks Torsion
Can we customize the assessment?

CONGENITAL HEART DISEASE

Insights into the Evolution of Myocardial Dysfunction in the Functionally Single Right Ventricle between Staged Palliations Using Speckle-Tracking Echocardiography

Edythe B. Tham, MBBS, Jeffery F. Smallhorn, MBBS, Sachie Kaneko, MD, Sabira Valiani, MD, Kimberley A. Myers, MD, Timothy M. Colen, MBBS, Shelby Kutty, MD, and Nee S. Khoo, MBChB, Edmonton and Calgary, Alberta, Canada; Omaha, Nebraska

<table>
<thead>
<tr>
<th>Stage</th>
<th>Mean age</th>
<th>(controls age)</th>
</tr>
</thead>
<tbody>
<tr>
<td>pre-Norwood</td>
<td>3.0 days</td>
<td>(4.0)</td>
</tr>
<tr>
<td>Inter-stage</td>
<td>5.3 months</td>
<td>(5.8)</td>
</tr>
<tr>
<td>Intermediate stage (Pre Fontan)</td>
<td>3.5 years</td>
<td>(3.8)</td>
</tr>
<tr>
<td>Post Fontan</td>
<td>3.7 years</td>
<td></td>
</tr>
</tbody>
</table>

Can we customize the assessment?

Speckle-Strain Assessment of the Single Ventricle

- End-systole
- Post-systolic shortening
- Neo-AVC
- Peak strain
- Q wave
No geometric assumption
Assess both longitudinal and circumferential strain
Strain rate is surrogate for contractility, relatively load independent
Dyssynchrony index
Post-systolic shortening
• No geometric assumption
• Assess both longitudinal and circumferential strain
• Strain rate is marker of contractility, relatively preload and HR independent
• Dyssynchrony index
• Post-systolic shortening

Schlangen et al. Circ Cardiovasc Img 2014
• No geometric assumption
• Assess both longitudinal and circumferential strain
• Strain rate is surrogate for contractility, relatively load independent
• Dyssynchrony index
• Post-systolic shortening

Speckle tracking echocardiography

Synchronous
Speckle tracking echocardiography

- No geometric assumption
- Assess both longitudinal and circumferential strain
- Strain rate is surrogate for contractility, relatively load independent
- Dyssynchrony index
- Post-systolic shortening a potential marker for tissue ischemia (coronary underperfusion)

Voight et al. JASE 2003
Can we customize the assessment?

**Significant Findings:**
Contractile indices change most in the interstage period.

**Longitudinal Strain:**
- Decreases 25% in the Interstage (pre-Glenn)

**Strain Rate:**
- Decreases by 50% in the Interstage. No change after volume unload
Can we customize the assessment?

**Significant Findings:**
Contractile indices change most in the inter-stage period.

**Circumferential Strain:**
preserved in the Inter-stage (pre-Glenn)

**Strain Rate:** Decreases by 50% in the Inter-stage
• Longitudinal study of 20 HLHS
  – Longitudinal strain
  – Circumferential strain
  – Longi/circ strain ratio

• Conventional measures
  – TAPSE, s’ velocity
  – RVFAC

Confirmed in 20 patients followed longitudinally
Does the RV adapt to a more LV-like pattern?

Pettersen et al. JACC 2007
Strain Rate as a predictor of Survival?

Longi SR > -0.97 1/s
AUC 0.82

Circ SR > -0.86 1/s
AUC 0.82

Lin, Khoo et al. AHA 2015
Birth to Inter-stage

- RVEF 70%
- RVEF 45%
- RVEF 32%

Synchronous

Dyssynchronous

Reducing Strain & Increasing dyssynchrony

This relationship appears confined to circumferential plane

Khoo et al. JACC imaging 2011
Friedberg et al. JASE 2007
Comparing Volume (MRI) and Dyssynchrony index

Circumferential mechanical dyssynchrony associated with RV dilation and hypertrophy
Pre Norwood to Post Fontan

Longitudinal MDI

Circumferential MDI

No significant change between stages suggest MDI plays a smaller role in ventricular changes between stages.
Case 2: HLHS Pre Norwood

RVFAC 40%
Normal TV annulus dimension

Why?
Why Image the Tricuspid Valve?

Pre Cavopulmonary anastamosis

RV FAC 31%
Aortic Arch obstruction (mild)
Pre Cavopulmonary anastamosis
Real-Time 3-Dimensional Echocardiography Provides New Insight Into Mechanisms of Tricuspid Valve Regurgitation in Patients With Hypoplastic Left Heart Syndrome

K. Takahashi, MD; A. Inage, MD; I. M. Rebeyka, MD; D. B. Ross, MD; R. B. Thompson, PhD; A. S. Mackie, MD; J. F. Smallhorn, MBBS

Conclusion—In hypoplastic left heart syndrome, moderate tricuspid regurgitation may be associated with increasing age, geometrical changes of the annulus, leaflet prolapse, lateral papillary muscle displacement, and subsequent leaflet tethering, as well as a smaller septal leaflet. (Circulation. 2009;120:1091-1098.)
Quantification of Tricuspid valve Annular & Leaflet Geometry (Tethering volume)

Circ Cardiovasc Imaging. 2014;7:765-772
Why Image the Tricuspid Valve?

Survival Associated with the degree of Tricuspid Valve Tethering

![Graph showing cumulative survival over follow-up months for low and high indexed tethering volumes. The graph indicates that survival is better for patients with low indexed tethering volume.](image)

Circ Cardiovasc Imaging. 2014;7:765-772
Is The repair for a Leaking TV always the same?

Posterior annuloplasty

Ohye J Thorac Cardiovasc Surg 2004
What do we learn from 3D imaging?

Pre Cavopulmonary connection
Annulus is now dilated (Z=3.5)

Leaflet Sizes: Normal
Commissures: Normal, no scallops or clefts
Adding the colour volume dataset
Central regurgitation
Extends to AL-SL closure line
Will a standard posterior annuloplasty address this?
Pre Fontan: FAC Now normal again (37%)

Moderate Central Regurgitation persists
Pre Fontan: determine the cause of TR

TV above
AL and SL are good size, appear to move well, not distorted

TV below
PL is smaller now (after annuloplasty repair)
Looking at the colour dataset

2 sources of regurgitation: part related to the PL (imbricated) is smaller.

Source of regurgitation not related to posterior leaflet annuloplasty
Is there leaflet prolapse or tethering?

From front of heart

PL rises higher into the plane of annulus

Tethering of the AL and the SL
Echocardiography: Why is it so useful?

2-Dimensional techniques/Flow and Colour Doppler
- End-Diastolic Area, End-systolic Area
- Calculated Fractional Area Change as a surrogate for ventricular volume determination.
- Regional wall motion abnormalities
- Qualitative information of many structures.
- Filling, ejection and isovolumic Intervals

Tissue Doppler imaging parameters
- Tricuspid annular plane systolic excursion of the RV free wall (TAPSE)
- Systolic tissue (S’) velocity
- Isovolumic acceleration (IVA) from color tissue Doppler: Force frequency response.
- Intervals: Myocardial performance index.

Speckle Tracking
- Strain and Strain Rate
- Dyssynchrony
- Post systolic shortening
- Directional motion, Torsion

3-Dimensional Echocardiography
- Volume determination, and ejection performance
- Complex anatomic structural detail (AV valves in particular)
Case 3

- **CASE 3**: 4 year old, Tricuspid Atresia, S/P Primary Glenn anastamosis at 3 months, Fontan at 2.5 years.
- Poor growth, persistent cyanosis, hepatomegaly ascites and pleural effusions,
- Being considered for
  - Fontan takedown
  - Surgical AV fistula
  - Transplantation
MRI demonstrates risk factors for poor survival after Fontan.

**FIBROSIS**

Late Gadolinium enhancement of the RV in patients with a systemic RV.

Rydman R et al, *Circ Cardiovasc Imaging* 2015
What are the long–term risks we should be aware of?

123 adult Fontan patients (22.4 years [±4.4]) post-Fontan

- Major adverse events occurred in 19 patients (15%).
- Transplant-free survival rates were
  - 95 %, at 20
  - 83 %, at 25
  - 60 %, at 30 years

- Modes of death
  - Fontan failure with preserved function
  - Congestive heart failure with decreased function
  - Sudden death
  - Thromboembolic event
  - Post-Fontan conversion
  - Post-transplant
What are the long–term risks we should be aware of?

Elder, Congenit Heart Dis. 2015;10:159–168

123 adult Fontan patients (22.4 years [±4.4]) post-Fontan

No differences in adverse outcomes were found based on morphology of the systemic ventricle, Fontan type, or systolic ventricular function.

Risk Factors

• Portal hypertension (OR 19.0, CI 4.7–77.3, P < .0001),
• Pacemaker (OR 13.4, CI 2.6–69.8, P = .002),
• Systemic oxygen desaturation (OR 0.86, CI 0.75–0.98, P = .02)
Hepatic Changes are not uncommon in the young adult

12 patients (average age 24 y) seen prior to extracardiac conversion of their Fontan.

- Reticular pattern of contrast enhancement in 8
- Zonal pattern of contrast enhancement in 4
- Hypervascular nodules in 2
- Multiple collateral vessels in 6 including GE varices in 4
- Hepatomegaly (10), splenomegaly (8), Ascites (2), Gall stones (4)

Kiesewetter et al, Heart 2007, Bryant Int J. Cardiol
Search for Biomarkers of Fibrosis and Hepatic dysfunction. Ultrasound or MRI elastography (Hepatic stiffness)

Kutty SS, JTCVS 2015


Type IV collagen level
FibroSURE (α2-macroglobulin, haptoglobin, apolipoprotein A1, bilirubin, γ-glutamyl transferase, alanine transaminase)
Echocardiography has developed a large repertoire of new tools.

- These are very well suited to assess complex questions related to contractile function, Valve function, dyssynchrony and other abnormalities.
- So far the predictive value of echo is limited to conventional measures, but this will change if prospective studies are funded.

MRI is best suited to answer questions related to the function of the Fontan circulation, and other organs:

- This is crucial for questions related to myocardial fibrosis,
- The impact of hepatic disease is significant, and severity is just beginning to be noted.
Conclusion
Thank you

• **Cardiovascular Surgery**
  Drs Ivan Rebeyka, David Ross, Mohammed Al-Aklabi

• **Echocardiography**
  Drs Jeffrey Smallhorn, Nee Khoo, Tim Colen, Shelby Kutty

• **MRI**
  Drs Edythe Tham, Dr Shi Joon Yoo (Toronto)
Can we customize the assessment of the RV?

Serial 3DE of RV volume

Kutty et al, JASE 2012
Surgical management

<table>
<thead>
<tr>
<th>Operative Techniques</th>
<th>Number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Annuloplasty</td>
<td>46 (85)</td>
</tr>
<tr>
<td>Commissuroplasty</td>
<td>29 (54)</td>
</tr>
<tr>
<td>Valvuloplasty</td>
<td>12 (22)</td>
</tr>
<tr>
<td>Chordal repair</td>
<td>2 (4)</td>
</tr>
<tr>
<td>Cleft closure</td>
<td>18 (33)</td>
</tr>
<tr>
<td>Edge-to-edge repair</td>
<td>17 (29)</td>
</tr>
<tr>
<td>Papillary muscle repair</td>
<td>1 (2)</td>
</tr>
</tbody>
</table>
Late hepatic complications after Fontan operation; non-invasive markers of hepatic fibrosis and risk factors

Jae Suk Baek,1 Eun Jung Bae,1 Jae Sung Ko,1 Gi Beom Kim,1 Bo Sang Kwon,1 Sang Yun Lee,1 Chung Il Noh,1 Eun-Ah Park,2 Whal Lee2

Seoul National University Children’s Hospital, Korea

Heart 2010;96:1750–1755.

$r=0.718$, $p<0.001$
Theoretical geometric advantage of circumferential contraction

Laplace’s Law

\[ \sigma = \frac{P \times R}{2h} \]

Apical 4CH

Basal Short axis
Impact of cardiopulmonary bypass

• Pediatric congenital heart surgery
• IVA a relatively load independent measure and atrial pacing
• Post CHD surgery reduction of contractility

Chung et al. JTCVS 2006
Can Imaging Make a Difference?

Can Imaging Differentiate?

- Compassionate Care
- Surgical Reconstruction
- Transplantation
Early Tricuspid Regurgitation in HLHS: mechanistic insights on tricuspid valve tethering and relationship with outcomes

Timothy Colen, Shelby Kutty, Richard Thompson, Chodchanok Vijarnspr, Ling Li, David Danford, Jeffrey Smallhorn, Nee Scze Khoo

(Longitudinal study)

Increased TV tethering and flatter bending angle prior to any surgical intervention is associated with TV regurgitation at medium term follow-up—prolapse was not present in these hearts at birth???

Accepted with revision-Circ CVS Imaging 2014
Pre Norwood to Post Fontan

More LV like pattern had better synchrony and RV ejection performance

Tham et al. JASE 2014
Pentality of Cantrell with a Single-Ventricle Cardiac Defect: Collaborative Management of a Complex Disease

Amy L. Marino • Richard J. Levy • John T. Berger • Mary T. Donofrio

Strengths:
Discern complex spatial relationships
• Topsy-turvy heart
• Arch and great vessel anomalies

Discern compression of / protrusion of Thoracic organs
• Diaphragmatic hernia
• Ectopia Cordis

Excellent resolution of non-moving structures.
• Great vessels

Dong et al. Journal of Cardiovascular Magnetic Resonance 2013, 15:40
Comparison Between Echocardiography and Cardiac Magnetic Resonance Imaging in Predicting Transplant-Free Survival After the Fontan Operation

127 patients, aged 12-23 with median followup of 3.8 yrs.
10% had an endpoint of death or transplant

Ghelani, S  Am J Cardiol 2015
<table>
<thead>
<tr>
<th>Decision Point</th>
<th>Question</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Fetal Life</strong></td>
<td>Will the RV or LV develop? (can intervention change this)</td>
</tr>
<tr>
<td></td>
<td>Is there a clear risk for non-viable newborn circulation?</td>
</tr>
<tr>
<td><strong>0-6 months</strong></td>
<td>Is there a possibility of Bi-ventricular repair?</td>
</tr>
<tr>
<td></td>
<td>Is the patient ready for a Cavopulmonary connection?</td>
</tr>
<tr>
<td><strong>2-3 years</strong></td>
<td>What are the risks/benefits of Fontan?</td>
</tr>
<tr>
<td><strong>3 years +</strong></td>
<td>Does the patient have Fontan Concept Failure?</td>
</tr>
<tr>
<td></td>
<td>Does the patient have Heart Failure?</td>
</tr>
</tbody>
</table>
Figure 2 | **Signal-to-noise ratio in clinical trials.** Detecting a statistically significant difference in outcome between treatment groups (for example, between experimental drug groups and placebo groups) can be conceptualized as a signal-to-noise problem. The higher the signal-to-noise ratio, the better the chances of detecting a true difference of a given magnitude and with a predefined sample size. **a** | The signal-to-noise ratio in a clinical trial with narrow patient selection criteria and a tightly controlled treatment scenario, such as clear definition of allowable concomitant medication and monitoring of patient adherence. This ‘clean’ setting is called an explanatory or efficacy trial, and the intergroup difference can be detected with ease. **b** | The signal-to-noise ratio of a pragmatic or effectiveness trial; in contrast to **a**, the trial population is more heterogeneous and the treatment scenario resembles everyday clinical practice. The signal may now be lost in the noise, even though the drug has not lost its pharmacological activity in those patients who are responsive and adherent; hence the signal is still there (orange oval) but is too small to detect.
Will the patient survive to a Fontan?

Impact of Initial Shunt Type on Cardiac Size and Function in Children With Single Right Ventricle Anomalies Before the Fontan Procedure

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<td>End-diastolic area, cm^{2}/BSA^{0.8}</td>
<td>284</td>
<td>17</td>
<td>1.15</td>
<td>&lt;0.001*</td>
</tr>
<tr>
<td>Transverse valve peak early velocity, m/s</td>
<td>300</td>
<td>17</td>
<td>4.46</td>
<td>0.08</td>
</tr>
<tr>
<td>Transverse valve peak atrial velocity, m/s</td>
<td>172</td>
<td>3</td>
<td>0.23</td>
<td>0.62</td>
</tr>
<tr>
<td>E/A ratio</td>
<td>172</td>
<td>3</td>
<td>1.78</td>
<td>0.54</td>
</tr>
<tr>
<td>Peak early diastolic velocity, cm/s</td>
<td>294</td>
<td>17</td>
<td>1.16</td>
<td>0.002*</td>
</tr>
<tr>
<td>Peak atrial diastolic velocity, cm/s</td>
<td>168</td>
<td>5</td>
<td>1.14</td>
<td>0.50</td>
</tr>
<tr>
<td>E/E' ratio</td>
<td>288</td>
<td>16</td>
<td>0.93</td>
<td>0.22</td>
</tr>
<tr>
<td>Flow reversal during atrial systole</td>
<td>291</td>
<td>17</td>
<td>0.29</td>
<td>0.05</td>
</tr>
<tr>
<td>MPI, inflow Doppler calculation</td>
<td>281</td>
<td>15</td>
<td>1.03</td>
<td>0.006*</td>
</tr>
<tr>
<td>Mitral diastolic flow velocity</td>
<td>281</td>
<td>15</td>
<td>2.42</td>
<td>0.68</td>
</tr>
</tbody>
</table>

Higher diastolic volume and inflow velocity is also associated with Death/Transplant

J Am Coll Cardiol 2014;64:2026–35
Anatomic variability may or may not be important. Non-HLHS patients do worse.
Will the patient survive to a Fontan/Overall?

Intermediate-term mortality and cardiac transplantation in infants with single-ventricle lesions: Risk factors and their interaction with shunt type

Having a Genetic Syndrome or Obstructed pulmonic veins is an extremely high risk situation
Will the patient survive to a Fontan/Overall?

Intermediate-term mortality and cardiac transplantation in infants with single-ventricle lesions: Risk factors and their interaction with shunt type

Socioeconomic status predicts survival
SV atrial function

• Minor enhancement of atrial compliance (reservoir function) can markedly increase cardiac performance
• 81 SV (mostly HLHS) vs. 51 controls
• Atrial distensibility (reservior), early diastolic passive flow (conduit), booster pump (active)
• Birth to post Fontan
• Conventional & deformation parameters

Khoo et al. J Am Soc Echocardiogr 2013
SV atrial function
Atrial strain

- Active strain
- Passive strain
- Reservoir strain
- MVC
- AVC
How does deformation correlate with Area change?

**Significant Findings:**
- Fractional Area Change decreases somewhat,
- Longitudinal and Circumferential strain may be measuring processes that are impacted differently by single ventricle physiology
- Correlation with fractional Area change is not exact.
• When is CMRI likely to fail

<table>
<thead>
<tr>
<th>Echo correct, CMR incorrect</th>
</tr>
</thead>
<tbody>
<tr>
<td>Two atroventricular canal defects (common atroventricular valve missed at CMR)</td>
</tr>
<tr>
<td>*Two TA (absent TV not clearly showed at CMR)</td>
</tr>
<tr>
<td>Ebstein anomaly (dilated right atrium diagnosed at CMR)</td>
</tr>
<tr>
<td>Two small VSD (3 mm, missed at CMR)</td>
</tr>
<tr>
<td>CoA (mild, missed at CMR)</td>
</tr>
<tr>
<td>PS (not clearly showed at CMR)</td>
</tr>
<tr>
<td>*Two right pulmonary artery hypoplasia (hypoplastic pulmonary artery not clearly showed at CMR)</td>
</tr>
<tr>
<td>TOF (overriding aorta not clearly showed at CMR)</td>
</tr>
</tbody>
</table>

Dong et al. Journal of Cardiovascular Magnetic Resonance 2013, 15:40
Pre Norwood to Post Fontan

Longitudinal PSSi

Circumferential PSSi

Increased PSSi at Pre BCPC

? Period of relative ischemia

Tham et al. JASE 2014
Questions

• What is the strength of correlation between observers and different platforms.
• Which system and Magnet do we use
Pre Norwood to Post Fontan

Reduction in SR suggest loss of myocardial contractility occurred between 1st interstage

Tham et al. JASE 2014
Risk Factors for poor Survival after Fontan

In a population of mixed systemic RV’s (post Fontan, & Mustard/Senning repairs, SPECT
• Perfusion scan identified moderate to severe perfusion abnormalities in 30%
• Reduced coronary reserve in the systemic RV (prone to ischemia).
• Progressive increase in EDP, decrease in Systolic function.

Hauser et al. Ped Cardiol 2003
Lubiszewska et al. JACC 2000
SV atrial function

- Single ventricle atrium is
  - Dilated
  - decr compliance,
  - decr early diastolic emptying
  - incr reliance on active contraction
- This deviates from normal early childhood maturational changes
- Appears to parallel atrium facing early ventricular diastolic dysfunction

Khoo et al. J Am Soc Echocardiogr 2013
Problem Solved?

• CASE 1: 4 month old, Univentricular connection with LAVV atresia, DORV.
• Urgent neonatal decompression of intact atrial septum
• Norwood Sano
• ECMO x 2
• Complex TV anatomy with tethering and straddle
• Being considered for Stage II. Has never left the hospital