Cardiac CT and MRI in Congenital Heart Disease

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Pediatric Cardiology
No Disclosures

• Thanks to Prakash Masand for CT images
Outline

• Common scenarios where CT and MRI are utilized

• How does CT and MRI add value in these patients
Trends in Cardiovascular CT/MR Imaging

Imaging Trends at Texas Children's Hospital

Data per month

Pediatrics
# CT versus MRI Choice

<table>
<thead>
<tr>
<th>MRI</th>
<th>CT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intracardiac anatomy, flow, function and viability imaging</td>
<td>✓</td>
</tr>
<tr>
<td>Comprehensive evaluation of extracardiac anatomy (heterotaxy, entire aorta)</td>
<td>✓</td>
</tr>
<tr>
<td>Definition of small vessels and higher order branches</td>
<td>✓</td>
</tr>
<tr>
<td>Emergent evaluation (especially neonates and infants)</td>
<td>✓</td>
</tr>
<tr>
<td>Airway, and lung parenchyma</td>
<td></td>
</tr>
</tbody>
</table>
CT Background

- Tremendous growth in Computed Tomography (CT) imaging
  - Fast scan times (0.27 to 0.35 seconds)
  - Excellent resolution (0.5 mm)
  - Physiologic information
  - 3-D post processing
Advantages: CT

• Complimentary to Echocardiography
• Fast acquisition with consistent results
• Excellent modality for emergent indications
• No sedation required
• Motion correction & Artifact reduction tools
Common Clinical Indications

• Complex anatomy in neonates & infants
• Coronary arteries
• Adult congenital heart disease with single targeted question
• Dynamic imaging
Evaluation of Pulmonary Arteries

- 2 day old for evaluation of branch pulmonary arteries
Evaluation of Pulmonary Veins

• Total anomalous pulmonary venous connection – Infracardiac type
Aortic Arch Assessment

- Severe aortic coarctation with tubular arch hypoplasia
Vascular Ring Investigation
Post Infantile Surgical Assessment

- Occluded Blalock Taussig Thomas shunt (BTT shunt)
Stage 1 Norwood: Sano shunt
Vascular stent
Coronary Artery Assessment

- Coronary ostial stenosis
Coronary Artery Assessment

Virtual Angioscopy
Coronary Artery Assessment
ACHD - RV-PA Conduit
ACHD - Ross procedure
ACHD – Atrial Baffle
Multiparametric Nature of Cardiac MRI

- **Morphology**: Are there anatomic anomalies?
- **Function**: Does the heart wall contract as it should?
- **Perfusion**: Is there sufficient blood supply to the heart muscle?
- **Viability**: Would the infarcted areas benefit from stenting?
- **Quantitative-Flow**: Are the valves leaking?
- **Coronaries**: Are there anomalous coronaries, or stenoses?
- **T1-mapping**: Is there diffuse fibrosis?
- **T2-mapping**: Is there edema?
- **T2*-mapping**: Is there iron overload?
TOF Predictors of Outcome

Electrophysiological Markers
- Prolonged QRS
- Sustained VT

Hemodynamic Sequelae
- RV Dilation
- Ventricular dysfunction
- Regional wall motion abnormalities

History
- Syncope
- Older age at repair

Geva T, JCMR, 2011
Impact of Pulmonary Regurgitation

Figure 3 Correlation between pulmonary right ventricular (RV) end-diastolic volume index and pulmonary pressure in patients with repaired TOF [46].

Figure 6 Factors influencing right ventricular (RV) dysfunction and impaired clinical status after TOF repair.

Geva T, JCMR, 2011
Post-Operative Echocardiogram
Echocardiography versus CMR

Margossian R, et al., Amer J of Cardiol, 2009
Post-Operative CMR
# Pulmonary Valve Replacement

<table>
<thead>
<tr>
<th>Study</th>
<th>RVEDVi</th>
<th>RVESVi</th>
<th>RV/LV</th>
<th>RV EF</th>
<th>LV EF</th>
<th>RVOTO</th>
<th>BPA Stenosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Geva T, et al</td>
<td>&gt;150 mL</td>
<td>&gt;80 mL</td>
<td>&gt;2</td>
<td>&lt;47%</td>
<td>&lt;55%</td>
<td></td>
<td>&gt;2/3rd Systemic</td>
</tr>
<tr>
<td>Oosterhof T, et al</td>
<td>&gt;160 mL</td>
<td>&gt;82 mL</td>
<td></td>
<td>&lt;45%</td>
<td></td>
<td></td>
<td>&gt;70:30% difference</td>
</tr>
<tr>
<td>Therrien, J, et al</td>
<td>&gt;170 mL</td>
<td>&gt;85 mL</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Knauth AL, et al</td>
<td>Z-score &gt;7</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Geva T, JCMR, 2011*
*Oosterhof T, Circulation, 2007*
*Therrien J, Amer J of Cardio., 2005*
*Knauth AL, Heart, 2006*
Branch Pulmonary Artery Stenosis

• Unilateral or bilateral branch pulmonary artery stenosis is a common complication
  - Maskatia, et al., 178 patients reviewed with 11% having bilateral stenosis, 26% unilateral stenosis
  - Harris, et al., 76 patients reviewed with 39% having either bilateral or unilateral stenosis

Maskatia S, et al., Amer J of Cardio., 2013
Harris MA, et al., JACC Imaging, 2011
Branch Pulmonary Artery Stenosis
Branch Pulmonary Artery Stenosis
Rapid Progression of RV Dysfunction

Wald R, et al., Heart, 2015
Protocol

- Axial Black Blood

- Cine imaging
  - VLA
  - 4 Chamber
  - SAX
  - RVOT
  - Branch PA’s

- Flow Quantification
  - Aorta
  - MPA
  - RPA / LPA
  - AVV

- 3D Whole Heart

- MRA

- Optional
  - Delayed enhancement if adult, history of depressed function or arrhythmia
D-TGA

• Second most common cyanotic heart lesion with incidence of approximately 30 / 100k live births
  - Approximate 5-7% of all CHD defects

• Approximately 40-45% have a VSD (Complex TGA)
  - Over half of these with concomitant LVOT obstruction

• Rare associated lesions include arch anomalies and pulmonary venous return abnormalities

Haeffele C, et al., Cardiol. Clinic, 2015
Villafane, et al., JACC, 2014
Arterial Switch Operation

- Majority survive to adulthood
  - Survival at 15 years ~ 88%
- Freedom from reintervention is 82% at 15 years
  - Reoperation < 10%

**TABLE 3** Post-Operative Sequelae Following the Arterial Switch Operation

<table>
<thead>
<tr>
<th>Long-Term Post-Operative Sequelae</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supravalvular pulmonary stenosis*</td>
<td>~10%</td>
</tr>
<tr>
<td>Supravalvular aortic stenosis*</td>
<td>~5%</td>
</tr>
<tr>
<td>Neoaortic root dilation</td>
<td>Nearly universal</td>
</tr>
<tr>
<td>Neoaortic regurgitation</td>
<td>Most (moderate or severe in &lt;10%)</td>
</tr>
<tr>
<td>Asymptomatic coronary occlusion</td>
<td>2%–7%</td>
</tr>
<tr>
<td>Sudden cardiac death</td>
<td>&lt;1%</td>
</tr>
<tr>
<td>Arrhythmia</td>
<td>2%–10%</td>
</tr>
<tr>
<td>Aortic dissection or rupture</td>
<td>Unknown</td>
</tr>
</tbody>
</table>

*Requiring intervention. Modified with permission from Wernovsky et al. (152).

Haeffele C, et al., Cardiol. Clinic, 2015
Villafane, et al., JACC, 2014
Coronary Ischemia

• Initial ASO mortality quite high due to coronary ischemia

• Incidence of myocardial ischemia is most prevalent in first 3 months
  - Bimodal pattern of incidence

• Coronary obstruction present in 5% - 7% of ASO survivors

Haeffele C, et al., Cardiol. Clinic, 2015
Villafane, et al., JACC, 2014
Coronary Ischemia After ASO
Protocol

• Axial Black Blood

• Cine imaging
  - VLA
  - 4 Chamber
  - SAX
  - RVOT
  - Branch PA’s

• Flow Quantification
  - Aorta
  - MPA
  - RPA / LPA
  - AVV

• 3D Whole Heart

• MRA

• Delayed Enhancement
Fontan
Decrease Exercise Tolerance

Complications of Fontan Palliation

Atrial and Ventricular Arrhythmia

Ventricular Systolic Dysfunction

Ventricular Diastolic Dysfunction

AV and Semilunar Valve

A-P and Venovenous Collaterals

Anderson, PA et al., JACC, 2008
Significance of Ventricular Dilation

Indexed volume > 125 mL

Ghelani S, et al., American J of Cardiology, 2015
Significance of Ventricular Dilation
Pulmonary Arteries

- Reliance upon unobstructed branch pulmonary arteries

**Table 6** Sensitivity, specificity, negative predictive value, positive predictive value, and accuracy of echocardiography to detect stenosis for each vessel segment

<table>
<thead>
<tr>
<th>Vessel</th>
<th>Sensitivity</th>
<th>Specificity</th>
<th>NPV</th>
<th>PPV</th>
<th>Accuracy</th>
</tr>
</thead>
<tbody>
<tr>
<td>RPA</td>
<td>67 (40–93)</td>
<td>65 (49–81)</td>
<td>85 (71–98)</td>
<td>40 (19–61)</td>
<td>42 (30–53)</td>
</tr>
<tr>
<td>LPA</td>
<td>54 (27–81)</td>
<td>72 (56–87)</td>
<td>79 (65–94)</td>
<td>44 (19–68)</td>
<td>42 (30–53)</td>
</tr>
</tbody>
</table>

**Figure 6** Scatter plots of vessel ratio measured using CMR and echocardiography: (A) scatter plot A, (B) scatter plot B.
A-P Collateral Quantification

A-P Collateral

Grosse-Wortman L, et al., Circulation: CV Imaging, 2008
Thrombus Evaluation
Ventricular Fibrosis
Protocol

- Gadovist with LOWER EXTREMITY IV
- Axial and Coronal Black Blood
- Cine
  - VLA
  - 4-Chamber
  - SAX
  - Axial Branch PA’s
  - Coronal PA’s
  - Aortic Arch
- Flow Quantification
- MRA
- Delayed Enhancement
Hypertrophic Cardiomyopathy

HCM

Fibrosis

Disarray
Diagnosis & Management
Diagnosis & Management
Risk Stratification in HCM

- Thickness of interventricular septum greater than 30 mm is an independent risk factor.
- Presence of delayed enhancement associated with arrhythmia and a 7-fold increase in adverse outcomes.

Maron M, et al., JCMR, 2012
Chan, et al., Circulation, 2014
Protocol

• Axial Black Blood

• Cine imaging
  - VLA
  - 4 Chamber
  - SAX
  - LVOT

• Flow Quantification
  - Aorta
  - MPA
  - AVV

• 3D Whole Heart

• MRA

• Delayed Enhancement

• Mapping Sequences
Lake Louis Criteria

Cardiovascular Magnetic Resonance in Myocarditis: A JACC White Paper

Matthias G. Friedrich, MD,* Udo Sechtem, MD,‡ Jeanette Schulz-Menger, MD,§
Godtfred Holmvang, MD,|| Pauline Alakija, MD,† Leslie T. Cooper, MD,¶ James A. White, MD,#
Hassan Abdel-Aty, MD,§ Matthias Gutberlet, MD, ** Sanjay Prasad, MD,††
Anthony Aletras, PHD,‡‡ Jean-Pierre Laissy, MD, §§ Ian Paterson, MD,|| ||
Neil G. Filipchuk, MD,* Andreas Kumar, MD,* Matthias Pauschinger, MD,¶¶
Peter Liu, MD,## for the International Consensus Group on Cardiovascular Magnetic Resonance
in Myocarditis
Lake Louise Criteria

- Lake Louise criteria comprised of increased signal on T2-weighted images, EGE ratio, and LGE
Lake Louise Criteria

- 13 centers, 143 patients, clinical diagnosis
  - LGE: 100% Abnormal in 81%
  - T2w: 69% Abnormal in 74%
  - EGE: 28% Abnormal in 55%
  - FPP: 48% Abnormal in 8%

- LV EDV, LVEF, RVEF predictive of persistent dysfunction

Banka P, et al., JCMR, 2015
Diagnosis & Management

[Image: Ultrasound scan with the following parameters: Freq.: 3.1 MHz/6.2 MHz, FPS: 90.1/]

Texas Children’s Hospital
Baylor College of Medicine
Myocarditis vs. DCM

- Does MRI have a predictive role in recently diagnosed DCM?

- Myocardial inflammation on CMR is a strong predictor of LV recovery:
  - 66 children within 2 weeks of DCM diagnosis
  - In 33/66, CMR identified at least one criterion of inflammation

Figure 4: Time to recovery of left ventricular function in CMR-positive group (full line) and in the CMR-negative group (dotted line) with 95% CI (grey dotted lines).

Raimondi F, Euro Heart Journal, 2015
Diagnosis & Management
Diagnosis & Management
CMR in All Myocarditis?

AHA/ACC Scientific Statement

Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 3: Hypertrophic Cardiomyopathy, Arrhythmogenic Right Ventricular Cardiomyopathy and Other Cardiomyopathies, and Myocarditis

A Scientific Statement From the American Heart Association and American College of Cardiology

Maron B, et al., Circulation, 2015
1. Before returning to competitive sports, athletes who initially present with an acute clinical syndrome consistent with myocarditis should undergo a resting echocardiogram, 24-hour Holter monitoring, and an exercise ECG no less than 3 to 6 months after the initial illness (Class I; Level of Evidence C).
It is reasonable that athletes resume training and competition if all of the following criteria are met (Class IIa; Level of Evidence C):

a. Ventricular systolic function has returned to the normal range.

b. Serum markers of myocardial injury, inflammation, and heart failure have normalized.
Additional Directions

- ACHD is a growing population
- Pulmonary Hypertension
- Interventional CMR
- Hematology & Oncology
Take-Home Points

• Cardiac CT allows rapid acquisition with typically low radiation doses and excellent spatial resolution

• Cardiac MRI has many strengths that extend beyond ventricular volume and function

• Myocardial characterization techniques are demonstrating value in non-CHD
Thank you....

• Cardiovascular Imaging team
• CT/MRI technologists and nursing
• Radiology colleagues