Pediatric CHD Imaging: What Adult Imagers should know

Rajesh Krishnamurthy,
Radiologist-in-Chief
Nationwide Children’s Hospital
Professor of Radiology
The Ohio State University
Columbus, OH

Disclosures
▪ Discuss the off-label use of ferumoxytol blood pool agent for MR angiography in children
▪ There is a FDA warning against the use of ferumoxytol as an intravenous contrast agent

Acknowledgements
▪ Cardiac Imaging Faculty at NCH: Drs. Kari Hor, Simon Lee, John Kovalchin, Julie O’Donovan, Steve Druhan, Cody Young, Eric Diaz
▪ Medical Imaging Scientists: Houchun ‘Harry’ Hu, PhD, and Ramkumar Krishnamurthy, PhD
▪ Technologists, analysts at the Pediatric Advanced Imaging Resource (PAIR) at NCH

Overview
▪ Selection
▪ Indications
▪ Modality
▪ Sedation, Acceleration and sequence optimization
▪ Standardization
▪ Acquisition
▪ Processing
▪ Interpretation
▪ Sharing
▪ Reporting Elements
▪ Showing value
▪ Quality
▪ Cost
▪ Outcomes
CHD is a team sport

- Clinical assessment
- Echocardiography
- Catheter Angiography
- Multidetector CT
- MRI

Conventional role of CT and MRI:
Augment echo and replace cath, where possible

<table>
<thead>
<tr>
<th>Echo</th>
<th>CT</th>
<th>MRI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Access</td>
<td>Access</td>
<td>Access</td>
</tr>
<tr>
<td>Coverage</td>
<td>Coverage</td>
<td>Coverage</td>
</tr>
<tr>
<td>Lumen</td>
<td>Lumen</td>
<td>Lumen</td>
</tr>
<tr>
<td>Wall</td>
<td>Wall</td>
<td>Wall</td>
</tr>
<tr>
<td>Flow</td>
<td>Flow</td>
<td>Flow</td>
</tr>
<tr>
<td>Operator</td>
<td>Operator</td>
<td>Operator</td>
</tr>
<tr>
<td>Time</td>
<td>Time</td>
<td>Time</td>
</tr>
<tr>
<td>Sedation</td>
<td>Sedation</td>
<td>Sedation</td>
</tr>
<tr>
<td>Radiation</td>
<td>Radiation</td>
<td>Radiation</td>
</tr>
</tbody>
</table>
Comprehensive evaluation of heterotaxy with MRI
Valvular Function and Flow

- Spatial resolution better than MRI
- Contraindications/Metal artifacts on MR
- Gold standard for non-invasive coronary imaging
- 3D-dataset with true isotropic resolution
- Dynamic Airway imaging

Alternative: CT

- Spatial resolution better than MRI
- Contraindications/Metal artifacts on MR
- Gold standard for non-invasive coronary imaging
- 3D-dataset with true isotropic resolution
- Dynamic Airway imaging
Hypoplastic left heart syndrome
s/p Norwood procedure and coiling of the aortic outflow
Assess pulmonary arteries, aortic arch and coronaries

Prospective EKG Gating with Target Mode

40% of cardiac cycle  28% of cardiac cycle
Effective doses significantly lower in target mode 320-detector group (0.51 + 0.19 mSv) compared to ungated 64-detector group (4.8 + 1.4 mSv), p<0.0001


Preoperative evaluation: Cardiac Morphology

- Atrial, ventricular and great arterial situs
- Segmental connections
- Status of the atrial and ventricular septum
- Cardiac valves
- Ventricular function
- Myocardium

- Echo is successful in delineating intra-cardiac pathology at all ages
- But, even in expert hands, some intra-cardiac defects remain difficult to diagnose by echo

Preoperative Evaluation: extra-cardiac vasculature

- Aortic coarctation
- Anomalous pulmonary veins
- Scimitar syndrome
- Systemic venous anomalies
- Branch pulmonary artery stenosis
- Anomalous coronary origin
- Coronary aneurysms

- Relatively high incidence of failure of echo to characterize extra-cardiac vascular pathology due to lack of acoustic windows
- Failure rate increases in the post-operative setting, and in older children, when acoustic windows diminish
Types of Coarctation

Diagnosis of CHD

Preoperative Evaluation

Morphology >>> Flow and function
CT = or > MRI (and diagnostic catheterization)

Postoperative Evaluation

Morphology well known
Evaluate function
Evaluate flow
Screen for complications
**Surveillance of Corrected CHD**

- Sequential measurements of RV volumes, mass and function in TOF, TGA
- Ventricular function after Fontan
- Surveillance of grafts, conduits and baffles
- Early detection of complications
- Determine timing of surgical intervention

---

**Tetralogy of Fallot Repair**

**Pulmonary regurgitation**

---

**RV Systolic Dysfunction**
RV Diastolic Dysfunction

Diagnosis of CHD
Post-operative Evaluation

Function and flow >>> Morphology
MRI >> CT

When is MRA a good choice
- Conditions requiring serial studies like coronary aneurysms in Kawasaki dz, aortic root size in Marfan, repaired coarctation, branch PA stenosis in TOF
- Screening studies
- Multiphasic studies
- Conditions requiring evaluation of flow, valvular, ventricular function and viability
- Renal insufficiency (using non-Gd techniques)
When is CT a good choice for CHD?

- Need for dynamic high resolution imaging. For e.g. coronary stenosis imaging
- Anomalous coronaries
- Vascular Rings and Slings
- Emergent situations like PE, trauma, aortic dissection, peri-graft leaks or occluded BT shunt
- Airway imaging
- Need to avoid sedation
- Metallic hardware

### Unrepaired TOF

<table>
<thead>
<tr>
<th>surveillance every 1-3 months in an infant before complete repair</th>
<th>TTE</th>
<th>TEE</th>
<th>MRI</th>
<th>CT</th>
<th>Stress</th>
<th>Angio</th>
</tr>
</thead>
<tbody>
<tr>
<td>surveillance every 1-6 months in an infant following valvuloplasty, PDA and/or RVOT stenting, or short placement before complete repair</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Evaluation after change in clinical status and/or new concerning signs or symptoms</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Evaluation to plan repair</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### TOF: after initial repair

<table>
<thead>
<tr>
<th>surveillance every 1-2 years in an asymptomatic patient with no residual sequelae or change in clinical status</th>
<th>TTE</th>
<th>TEE</th>
<th>MRI</th>
<th>CT</th>
<th>Stress</th>
<th>Angio</th>
</tr>
</thead>
<tbody>
<tr>
<td>surveillance every 1-2 years in an asymptomatic patient with no residual sequelae or change in clinical status</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Evaluation every 6-12 months in a patient with residual right ventricular outflow tract obstruction, stenosis, or atrioventricular septal defect</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Evaluation every 3-12 months in a patient with heart failure symptoms</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Surveillance every 3-5 years in a patient with pulmonary regurgitation and preserved ventricular function</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre-procedural evaluation before pulmonary valve replacement (percutaneous or surgical) including evaluation of the proximal courses of the coronary arteries</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Sedation, Acceleration and Sequence Optimization

Delayed Risks associated with procedural sedation

- Potential risk of neurotoxicity:
  - Impairment of memory and recollection later in life after GA in infancy: independent of underlying dz ¹
  - Developmental and behavioral disorders in children who had surgery younger than 3 years is 60% greater than siblings without surgery ²

Reducing Sedation for Pediatric Imaging

Imaging Pathways for reducing sedation risks

- Distraction, noise-reduction, and feed and wrap techniques
- Use of alternatives to sedated MRI: ultrasound, low dose, unsedated and free-breathing CT
- Advanced motion correction schemes for respiratory, cardiac and gross patient motion
- Avoid GA and convert protocols to intravenously sedated free-breathing acquisitions
- Achieve protocol brevity by use of targeted sequences that are proven to change management and influence outcomes
- Achieve protocol brevity by use of a few targeted 3-D sequences rather than multiplanar 2D

Combining Strategies: Preserving Spatial Resolution

Parallel Imaging Techniques (SENSE) ➔ 2x-4x
+ Partial Fourier Acquisitions ➔ 2x-3x
+ Temporal sub-sampling strategies ➔ 4x–5x
Sub-second CE-MRA ➔ 16x–60x

Time resolved 4D MRA
Practical Clinical Utility: Adapting the MRA to different clinical scenarios

- Improve spatial resolution in larger patients with slower circulations along with preserved separation of phases
- Achieve better temporal resolution (e.g. neonate with rapid circulation to achieve separation of phases)
- Dual injection: Split the dose of Gd between extremities
- Decrease contrast dose, but preserve signal
- MRA with non-phased array coils (without SENSE), to reduce dynamic time
- First pass venography

Improve temporal and spatial resolution, intubated

Newborn with heterotaxy, obstructed supracardiac TAPVR, 135 bpm, flex M coil, keyhole, parallel imaging, BH, 2.4 sec dynamic, ref scan 9.6 sec, 2 cc/sec injection with saline flush

Free breathing, needs separation of R and L sided structures with good spatial resolution

14 mo old M, William’s syndrome, Supravalvular AS
5 second dynamic with parallel imaging
First Pass venography
Systemic vein thrombosis after arterial switch procedure, 1 y M, free breathing
Dilution of contrast, keyhole, parallel imaging, 2 sec dynamic

2 separate injections into the upper and lower extremity in Fontan
Choice of Contrast Agent

• Conventional extracellular agent (e.g. Gadopentetate, Gadoterate, Gadobutrol etc.)
• Extracellular-Intracellular (e.g. Gadoxetate)
• Blood pool contrast agent (Ferumoxytol)

Steady state free breathing MRA with slow injection protocol of extracellular contrast

• Navigator respiratory gated IR prepped 3D TFE

Free breathing, HR 105 bpm; Scan time ~ 5 min; 1 mm isotropic resolution

Golden-Angle Ordering Scheme

• Angle continuously increased by 111.25°
• Never acquires same spoke twice
• Next spoke fills largest angular gap
• Spokes always cover k-space uniformly

• Optimal distribution of spokes → Reconstruction from arbitrary windows
• Retrospective selection of temporal resolution

Simplification of Clinical Workflow

- No timing / synchronization failures → phases never missed
- No estimation of circulation delay needed → no test bolus
- No voice commands needed → no language barrier
- No patient cooperation required → elderly / sick / pediatric
- Previously difficult exam → "One-click" procedure

Morphologic analysis using cine 3D SSFP
5 yo F, Heterotaxy, unbalanced CAVC, s/p DKS, arch repair, separate MV insertion into common atrium, pre Fontan evaluation

Integrated 3D cine SSFP and navigator 4D flow
Why native 3D Imaging in CHD?

- Comprehensive, customized clinical imaging
- Reduces chances for patient recall
- Measurements customized to every patient
- Unsupervised imaging
  - Reduced operator dependence
  - Remote locations
  - Reduces image registration issues
  - Standardization
- Can address unknown/complex questions
  - Collateralization in Fontan circulation

40 y f, Dextrocardia, (I,L,D) transposition of great arteries, large unrepaired ASD, mild PS

2D PC MPA
Reconstructed outflow tract 3D PC

2D PC AvV
Reconstructed AV inflow 3D PC

Protocol for FB 3D imaging of CHD

< 1 min
2–4 min
5–7 min
4.5–7 min
6–12 min
Results: Comparison of volumetry and flow between 2D and 3D

Figure 2: Plots showing results from 2D and 3D cine and flow imaging. (A) shows the 2D vs. 3D comparison between volumes (mL). A significant correlation is seen indicating the clinical utility of 3D cine imaging. (B) shows the stroke volume measured across different flow structures (aorta, Ao, MPA) in the aortic root. A broader correlation between 3D and 2D flow is seen here.

Vessel Diameter

- 3D: 24 mm (95% CI 20-28)
- 2D: 23 mm (95% CI 19-27)
- MRA: 21 mm (95% CI 18-25)
- 3D: 20 mm (95% CI 17-22)
- MRA: 22 mm (95% CI 19-25)

Results: Qualitative Analysis

- ISA 9% better on 3D vs 2D
- 3D scores of tricuspid & aortic valves 21-29% lower than 2D
- 3D scores 23-57% lower than 2D score for 1st/2nd order branches, AV separation
- 3D scores 25% lower for veins
4D Flow

Standardization

What is needed to scale for impact?

Standardize, standardize, standardize...

- Standardized operating protocols
- Standardized imaging protocols
- Standardized reporting
- Standardized metrics
- Integrated HIS/RIS/reporting/research database
**Challenges**
- Standardization
  - Across various vendors
    - Think about 2D phase contrast imaging
  - Different acceleration and acquisition techniques
    - Compressed sensing
    - Non-cartesian imaging techniques
    - Reconstruction time
  - Post processing techniques/ methodology
- Contrast enhanced imaging
  - Post ablavar, pre ferumoxylon era

**Integration of Imaging Data into the healthcare enterprise (IHE)**
Standards allow exchange of clinical/imaging data:
- HL7 v3
- Cross-enterprise document sharing (XDS, XDS-I)
- FHIR (Fast Health Interoperability Resources)
  - *Enrich patients’ clinical record*
  - *Provide reliable, authorized access to it across the enterprise (and beyond)*

**Sharing and Showing Value**
*Reporting Elements Registries*
Virtual angioscopy (left) and intraoperative photography (right) of anomalous RCA with slit-like ostium

Arrow: LCA
Arrowhead: Slit like ostium of RCA

Sudden Cardiac Death in the Young
AAOCA: Key Reporting Elements

A. Type of AAOCA
B. Ostial morphology
C. Location of coronary ostia
D. Ostial relationship
E. Presence and length of intramurality
F. Course through commissure or column
G. Dominance

Screening/diagnostic tools in SCD

• Who is at risk of sudden death?
• What is the relative risk of anomalous R. vs L.?
• What are the morphological factors associated with increased risk?
• How good are we at diagnosing/grading risk factors?
• How do we decide management: observation, exercise restriction, surgery?
Arterial tortuosity and outcomes in Marfan syndrome and Loeys-Dietz syndrome

Increased Vertebral Artery Tortuosity Index Is Associated With Adverse Outcomes in Children and Young Adults With Connective Tissue Disorders
Shane A. Morris, MD; Dennis B. Goehel, MD, PhD; Tai Geva, MD; Michael N. Singh, MD; Kimberlee Guerra, ScD; Ronald V. Lauro, MD

Freedom from Thoracic Aortic Dissection – Marfan syndrome

Imaging Metrics in ACOs: Looking beyond Process Metrics

• By contributing directly to improved patient outcomes
  – Reduced mortality from sudden cardiac death in children based on the provision of effective screening programs
  – Reduced number of imaging-related morbidity related to sedation and radiation

• By contributing to reducing all costs over an episode of care
  – Accurate and timely diagnosis of neonatal manifestations of CHD not adequately characterized by echo, which results in reduced need for catheterization, duration of surgery, and intra-operative manipulation/imaging.
  – Prompt access to specialist imaging services, which reduces length of stay in the ED and hospital

• By contributing to reducing the length of an episode of care
  – Reducing time to diagnosis with
    • Early imaging or imaging-guided intervention
    • Time to initiation of treatment
    • Determination of short-term response to therapy
Reducing need for sedation for chest/cardiovascular imaging in little children aged 0-4 years

Reducing radiation from cardiovascular CT scans in Children

‘DORV in your Hands’ Workshop
Looking to the future...

• Pediatric CV imaging field at risk from impending changes in reimbursement
• Organized and data-driven attempt to demonstrate value of imaging in discrete patient care pathways requires a quality focus
• Broad user support in the community for shared data collection for critical questions
• Reduce shotgun approach to imaging utilization
• Patient-centric focus brings stakeholders together

17th SPR Hands-On Course on Pediatric Cardiovascular MR and 15th SPR Advanced Symposium on Pediatric Cardiovascular Imaging
October 15 – 23, 2019 | Nationwide Children's Hospital
Columbus, Ohio

This year, the SPR CV meeting will be held in conjunction with the 3D Imaging for Interventional Cardiomechanics in CHD (3DCI).

Join us for a unique joint conference combining The Society for Pediatric Radiology (SPR) and the International Symposium on 3D Imaging for Interventional Cardiomechanics in CHD (3DCI). Co-organized by cardiac imaging radiologists Dr. Rajesh Krishnamurthy, cardiologist Dr. Ken Hor and interventional cardiologist Dr. Almas Ammouni, this combined conference will feature interactive hands-on training.

VISIT SP Rcours Nationwid Childred.org to learn more or to register.