Acquired Heart Disease in Adults with Congenital Heart Disease: What to Image? How to Image? When to Image?

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SPR 2015
Congenital cardiovascular disease (CHD)

- Affect at least 1% of newborn infants
- Patients born with CHD survive into adulthood
- Advances in pediatric cardiology and cardiac surgery
- 3 million population in US with CHD at least half have complex defects
- Adults with CHD (ACHD)
  - palliative or reparative surgery during early life
  - life-long follow-up
Transition of care multidisciplinary pediatric and adult teams

Cardiology
Cardiac surgery
Cardiac anesthesia
Cardiac perfusion
Cardiac imaging
Critical Care

Adult Congenital Heart Disease

Hepatology
Nephrology
Hematology
Pulmonary Hypertension
Maternal/Fetal Medicine
Heart Failure & Transplantation
Adult CHD – disease pattern

- **Primary – Unrecognized or undiagnosed during childhood**
  - Aortopathies
  - ASD/VSD
  - Partial anomalous pulmonary venous return

- **CHD defects – underwent corrective/ palliative surgery during childhood**
  - Residual or recurrent lesion – medical, surgical, interventional treatment in adult life

- **Special needs patients with associated CHD**
  - Down; Velo-cardio-facial; Turner; Noonan Syndromes

- **Acquired heart disease**
  - coronary artery disease
  - arrhythmias,
  - heart failure
Imaging ACHD - Echocardiography

**Methodological strengths**
- 1st line diagnostic modality in ACHD
- Real time acquisition – no ionizing radiation
- Portable
- Local velocity measurements
- M-mode traces and 3D
- Suitable for OR

**Clinical strengths**
- Applicable in almost all cases
- Well visualization
  - valve disease
  - jet velocities
  - septal defects
  - vegetation

**Clinical weakness**
- Limited visualization
- Right ventricle
- Pulmonary arteries
- Pulmonary veins
- Distal ascending aorta/ transverse arch

**Methodological limitations**
- Limited windows, angles and depth of access
- Not suitable for volumetric flow measurements
Imaging ACHD - MRI

**Methodological strengths**
- Not restricted by body size or poor acoustic windows
- Non ionizing radiation
- Multi-slice cine coverage
- Biventricular function
- Volumetric flow measurements
- Tissue characterization
- Dynamic angiography

**Methodological limitations**
- Image slice thickness
- Time consuming acquisition
- Expensive
- Pacemakers are regarded as contraindication

**Clinical strengths**
- Aortopathies
- Tetralogy of Fallot
- Shunts
- TGA
- Single ventricle physiology - Fontan procedure

**Clinical weaknesses**
- PFO or small ASD
- Mobile vegetation
- Measure tricuspid regurgitation and jet velocity for RV pressure estimation
Imaging ACHD - CT

Methodological strengths
- Excellent spatial resolution
- 3D visualization
- Short acquisition time
- Calcium visualized well
- Vascular lumen generally seen within stents

Methodological limitations
- Ionizing radiation
- No flow measurements
- Limited cine visualization

Clinical strengths
- Coronary arteries and collaterals
- Conduits and stents
- Associated lung or upper abdominal pathology

Clinical weaknesses
- Limited functional information
- Unsuit for long term followup due to radiation
ACHD - Unrecognized or undiagnosed during childhood

- ASD/VSD - echocardiography modality of choice
- MRI
  - Clarifies the nature of lesion
  - Qp:Qs
  - Biventricular size and function
  - Associated anomalies – PAPVR

- PDA – MRI/CT
  - Visualize the defect
  - Qp:Qs
Coarctation /Recoarctation

- MRI/CT
  - De novo diagnosis
  - Arterial collaterals
  - Flow patterns
  - Associated aortic valve anomalies
  - Aortic dimensions
  - Biventricular function
  - Evaluation for possible stent aortoplasty- arch geometry

- Post surgical repair MRI –Clinical suspicion for recoarctation
  - Recoarctation/Restenosis
  - Complications- Aneurysm formation at repair size
  - Aortic valve stenosis/ regurgitation
  - Aortic dimensions
  - LV hypertrophy
  - Post stent aortoplasty follow-up CT in 3 mo
Aortopathies

- MRI/CT
  - Aortic valve anatomy and function
  - Aorta
    - Dimensions
    - Aneurysmal dilation
    - Dissection
  - Flow patterns
    - 4D Flow
  - LV hypertrophy
  - Biventricular function
Tetralogy of Fallot - MRI

- Location and severity
  - RVOT obstruction,
  - pulmonary artery obstruction
- Branch PAs flow and distribution
- Pulmonary regurgitation
- Atrial and ventricular residual septal defects
- Qp/Qs
- RV/LV volumes and ejection fraction
Complex repaired CHD – MRI/CT

**D-TGA atrial switch**
- Assess atrial pathways
  - Severity of obstruction
  - Baffle leaks - not well assessed
- RV & LV size and function
- RVOT/LVOT obstruction mechanisms
- Qp:Qs evaluation
- Tricuspid regurgitation
- Systemic RV size and function
- Branch PAs flow distribution
- SVC/IVC flow ratio

**D-TGA arterial switch**
- RVOT, supravalvar PA stenosis, branch PA stenosis
  - Measurements
  - Flow
- Neo-Aortic valve
- Biventricular function
- Coronary assessment
- Myocardial fibrosis/ ischemia
Single ventricle Fontan – MRI/CT

- Cavopulmonary/atriopulmonary connections
- Branch pulmonary arteries
- Pulmonary veins: narrowing/stenosis
- Aortic arch obstruction
- Ventricle size and function
- Atrioventricular valve
- Ventricular outflow tract
- Aortopulmonary collaterals

- Complications:
  - Arrhythmias
  - Thromboemboli – hypocontractile right atrium
  - Protein losing enteropathy (PLE)
  - Hepatic dysfunction
  - Systemic venous collaterals
  - Pulmonary arteriovenous malformations (AVM)
Acquired heart disease

- Ischemic coronary artery disease
  - Anomalous coronary origins
  - Re-implanted coronary arteries in complex CHD
  - Kawasaki disease
  - Acquired atheromatous disease superimposed on CHD
- Arrhythmias
  - Multiple open heart surgeries
  - Tunnels, baffles
  - ASD/VSD closure
- Heart failure
  - Right ventricular dysfunction
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