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Adult Congenital Heart Disease

Saurabh Rajpal, MBBS, MD
Assistant Professor

Approach to CMR in ACHD
- Diagnosis/Op Reports
- Clinical Questions
- Protocol
- Interpretation
- Report

Today’s Talk
- Segmental Anatomy
- Common CHD Diagnosis/Complications
- Protocol Design
- Interpretation
- Reporting
Segmental Anatomy

- Where are the thoracoabdominal viscera? (S)(I)(A)
- Where is the heart positioned? (Dx)(Lx)
- Atrial situs? (S,I,A)
- How are the ventricles looped? (D)(L)

Segmental Analysis

- How are the ventricles looped? (D)(L)
- Are the AV connections ok? (Con)(Disc)(Str)
- How do the ventricles align to the great arteries?
- Relationship b/w the semilunar valves? (S)(D)(L)

{S,D,S}

Atrial Situs
Ventricular Loop
Semilunar Valve Roots
A Very Simplified Example

(S,O,D) Transposition of Great Arteries

D- Loop Transposition of Great Arteries

Tetralogy of Fallot

- Most common cyanotic CHD
- >90% survive to adulthood
- Surgery in first year of life
- Palliative shunts
- Surgical goals
  - Relief of RVOT Obs (+/- Vtomy)
  - Pulm Valvotomy
  - Musc bundle resection
  - TAP
  - RV-PA conduit
  - Separation of circulations
    - VSD Patch

Repaired Tetralogy of Fallot in Adults

- Pulmonary Valve Regurgitation
- Tricuspid Valve Regurgitation
- Residual VSD
- Residual ASD
- RVOT Aneurysm
- AP Collaterals/Shunts
- RV Fibrosis
- Dilated Aorta
- LV Dysfunction
- Coronaries (TPVR)
- Branch PA stenosis
- Arrhythmias/SCD

CMR in rTOF
Gold standard for follow-up of RV size and function and PR
Generally repeated every 12-36 months
Pulmonary Valve Regurgitation

RV Function

Quantification of Pulmonary Regurgitation

Phase Contrast Imaging

- PC Imaging
- PR Fraction by PC Imaging
- PR = Pulm Outflow - Ao outflow
- Cine SSFP
  - PR = RVSV - LVSV

Valente et al. JASE 2014

Indications for Pulmonary Valve Replacement

- PR fraction >25% and
  - Either
    - Symptoms
    - Or (at least 2)
      - Mild or greater RV or LV dysfunction
      - Severe RV dilation
      - RV EDV index ≥160 mL/m2,
      - RV ESV index ≥80 mL/m2
      - RV EDV ≥2 times the LV EDV

Stout KK et al. JACC 2018
rTOF- Patterns of LGE

Adult rTOF Protocol
- Cine CMR
  - Ventriculography
  - RVOT long axis
  - Axial stack branch PAs
- PC
  - Aoa
  - MPA
  - Branch MPA
- CE-MRA or 3D SSFP
- LGE

Adult rTOF Reporting Elements
- RV/LV Function
- PR Fraction
- RVOT obst (location and severity)
- Branch pulmonary artery flow distribution
- Qp/Qs
Transposition of Great Arteries
Ventriculoarterial discordant lesion
Aorta → RV → PA → LV

D loop TGA - VA Discordance
L loop TGA - VA and AV Discordance

Surgical Repairs - Transposition of Great Arteries

Atrial switch
Arterial switch
Rastelli procedure

TGA - Atrial Switch
- Systemic baffle stenosis/leak
- Pulmonary baffle stenosis/leak
- Coronaries (compression)
- Systemic RV function
- Sub-pulmonic LV function
- Valve regurgitation
- RV/LV Fibrosis
- Qp/Qs for shunt fraction
TGA - Arterial Switch
- RV outflow tract stenosis
- Branch PA stenosis
- Neo-aortic root dilation
- Neo-aortic valve regurgitation
- Re-implanted coronary artery assessment
- Ventricular function
- Stress CMR
- Valve regurgitation assessment
- LGE- Viability

Congenitally Corrected TGA or L-TGA
- TR (systemic)
- Systemic RV Dysfn
- Complete Heart Block
- Asstd - VSD and PS

Coarctation of Aorta
- Narrowing of the descending aorta
- Hypertension
- Intracranial aneurysms
- Aortic aneurysm
- Aortic dissection
- Spinal subarachnoid hemorrhage
- Collaterals
- Premature CAD
- Stroke
- Surgery, Balloon Angioplasty, Stent
Coarctation of Aorta – Protocol

- Cine CMR:
  - long-axis aortic arch
  - short-axis aortic root
  - ventriculography
- CE-MRA or 3D SSFP
- PC CMR: collateral flow
- LGE

Flow at Diaphragm - Flow at CoA = Collateral Flow

Coarctation of Aorta - Report

- Coarctation
  - location
  - dimensions
  - severity
- Arch sidedness and branching
- Aneurysm
- Dissection
- Collateral vessels to the DAo
- Ventricular parameters
- Associated valve disease

Coarctation of Aorta - 3D SPACE TSE
Shunt Lesions

- ASD - RA/RV Dilation
- VSD - LA/LV Dilation
- PDA - LA/LV Dilation
- Gerbode Defect - 4 Chamber Dilation

Quantify the Shunt: Qp/Qs

Qp/Qs = Ratio of RV and LV Stroke Volumes

Gerbode Defect
RV Dilation

Tricuspid regurgitation
Pulmonary regurgitation
Pulmonary artery hypertension
Shunt Lesions
Myocardial abnormalities
- Uhl's anomaly
- ARVC
- Ventricular dysfunction

Etiology of RV Dilation

Planning - Oblique Sagittal Atrial Septum
Ebstein Anomaly

- Abnormal TV and RV
- Tricuspid regurgitation
- RV Failure
- ASD - O2 desaturation
- Atrial Arrhythmias
- Surgery- TVR or Cone procedure
Ebstein Anomaly

Complex Congenital Heart Disease
Thank you