Prognostic imaging in congenital heart disease

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Disclosures

- No financial disclosures
CT and MRI as prognostic tools

- **Aorta**
  - Congenital aortic stenosis/insufficiency
  - Aorta coarctation

- **Right ventricle size**
  - Anomalies of the pulmonary artery or valve
  - Tetralogy of Fallot or Truncus arteriosus

- **Complex palliations**
  - Univentricular heart
  - Hypoplastic left heart syndrome
Ascending aorta aneurysm

- Congenital aortic stenosis/insufficiency
- Collagen vascular disease

41 year old woman with Marfan disease

18 year old man with bicuspid aortic valve
Marfan syndrome

- Prophylactic aorta root replacement 5.0 cm
- Aorta diameter < 5.0 cm
  - Rapid growth > 1.0 cm/year
  - Family history premature aortic dissection
  - Moderate or greater aortic regurgitation
- MRI or CT
  - Annual if abnormal beyond aorta root
  - 5 years if normal beyond aorta root


Bicuspid aortic valve progression of aorta root and ascending aorta dilatation

- Diameter ↑ up to 2 mm/year
- Annual follow up recommended:
  - Aorta dilatation > 4.5 cm
  - Rapid change in aorta diameter
  - + family history of aortic dissection
- Longer interval in others

Bicuspid surgery practice

- **Valve surgery** → 61% replace aorta if diameter 4.5 cm
- **No valve surgery** → 55% replace aorta if diameter 5.0 cm

20 year old with bicuspid aortic valve & ascending aorta aneurysm

Coarctation of the aorta

42 year old with history of coarctation repair
Aneurysm formation following coarctation repair

Associations
- Bicuspid aortic valve
- Patient age

- No BAV and age < 20 years 3%
- BAV and age > 39 years 50%

Coarctation complication detection

- 50% re-coarctation cases: no clinical findings
- MRI re-coarctation detection rate same for symptomatic & asymptomatic patients
- Aorta aneurysms are not detected by echo


Lifelong cardiology follow-up is recommended for all patients with aortic coarctation (repaired or not), including an evaluation by or consultation with a cardiologist with expertise in ACHD.

Evaluation of the coarctation repair site by MRI/CT should be performed at intervals of 5 years or less, depending on the specific anatomic findings before and after repair.

Right ventricle size

- Anomalies of the pulmonary artery and valve
- Tetralogy of Fallot and Truncus arteriosus
- Transposition complex
Valved pulmonary artery replacement

- Pulmonary regurgitant fraction $\geq 25\%$
- Right ventricle enlargement
- Abnormal cardiac function
- QRS duration $> 140$ msec
- Associated anatomic abnormalities

Geva T. Repaired tetralogy of Fallot: the roles of cardiovascular magnetic resonance in evaluating pathophysiology and for pulmonary valve replacement decision support. Journal of Cardiovascular Magnetic Resonance 2011; 13: 9
Measures of cardiac function

- **Systolic function**
  - RV ejection fraction < 47%
  - LV ejection fraction < 55%

- **Diastolic dysfunction**
  - Abnormal tricuspid flow patterns
  - Late diastolic forward flow

Geva T. Repaired tetralogy of Fallot: the roles of cardiovascular magnetic resonance in evaluating pathophysiology and for pulmonary valve replacement decision support. Journal of Cardiovascular Magnetic Resonance 2011; 13: 9
Right ventricle end diastolic volume index

- RV EDVI > 150 ml/m² (z-score > 4)
- Right ventricle remodeling threshold limit
Complex palliations

- Univentricular heart
- Hypoplastic left heart syndrome
Ventricle size and prognosis

Right ventricle ejection fraction 38%
Right ventricle end diastolic volume 265 ml
RV end diastolic volume index 156 ml/m²

Kaplan-Meier plot of cumulative freedom from the end point (death or transplantation stratified by indexed end-diastolic volume (EDV) > 125 ml/BSA^{1.3} (P<0.001)

Rathod RH, et al. Cardiac magnetic resonance parameters predict transplantation-free survival in patients with Fontan circulation.
Summary: key prognostic indicators

- Aorta aneurysm: MRI and CT diameter measurements
- Right ventricle regurgitation: RV EDVI for valve replacement
- Univentricular heart palliation: RV EDVI for survival prognosis