Role of imaging in Heart Failure: Heart failure in corrected congenital heart disease

Sotiria C. Apostolopoulos

Dept of Pediatric Cardiology & Adult Congenital Heart Disease

Onassis Cardiac Surgery Center
Imaging modalities

- Chest X-Ray
- Echocardiography
- Cardiovascular Magnetic Resonance
- Cardiovascular Computed Tomography
- Nuclear Scintigraphy
- Barium Swallow
- X-ray Angiography
Tetralogy of Fallot - Repair

TETRALOGY OF FALLOT, REPAIRED

YSD Patch Closure
Infundibular Resection
RV Outflow Patch

TOF Repair

↑ Pulmonary regurgitation

PA dilation
(↑ capacitance)

RV dilatation

↑ PA pulse pressure/volume

↑ RV compliance

↑ Pulmonary regurgitation
Table 3: Echocardiographic reference values of RA and ventricular size and function in healthy adults (adapted from Rudski et al.24)

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Abnormal</th>
</tr>
</thead>
<tbody>
<tr>
<td>RV diameter (mm)</td>
<td>&gt;42</td>
</tr>
<tr>
<td>Base</td>
<td></td>
</tr>
<tr>
<td>Midventricular level</td>
<td>&gt;35</td>
</tr>
<tr>
<td>RV diastolic length (mm)</td>
<td>&gt;86</td>
</tr>
<tr>
<td>RV diaphragmatic wall thickness (subcostal view) (mm)</td>
<td>&gt;5</td>
</tr>
<tr>
<td>Systolic function</td>
<td></td>
</tr>
<tr>
<td>TAPSE (mm)</td>
<td>&lt;16</td>
</tr>
<tr>
<td>Pulsed Doppler peak S' (cm/sec)</td>
<td>&lt;10</td>
</tr>
<tr>
<td>FAC (%)</td>
<td>&lt;35</td>
</tr>
<tr>
<td>Diastolic function</td>
<td></td>
</tr>
<tr>
<td>E/E' ratio</td>
<td>&gt;6</td>
</tr>
<tr>
<td>RA end-systolic area (cm²)</td>
<td>&gt;18</td>
</tr>
<tr>
<td>RA length (base to apex) (mm)</td>
<td>&gt;53</td>
</tr>
<tr>
<td>RA lateral diameter (mm)</td>
<td>&gt;44</td>
</tr>
</tbody>
</table>

FAC, Fractional area change.
Repaired TOF: PI - TR

Mild PI: persistent flow gradient at end-diastole
Moderate PI: equilibration of MPA and RV pressures only at end-diastole
Severe PI: early diastolic pressure equilibration

LPA PW showing degrees of diastolic flow reversal

TR morphology
RVp estimate = $4v^2 + RAP$

J Am Soc Echocardiogr 2014;27:111-41
Repaired TOF: RV

RV fractional area change (%)
(modest correlation with CMR in TOF)

TAPSE
(weak correlation with CMR in TOF)

J Am Soc Echocardiogr
2014;27:111-41
**Repaired TOF: Newer techniques**

- **2D Speckle tracking**
  - (no available guidelines in CHD)
  - Echocardiography. 2014;31(4)

- **TDI**
  - (no available guidelines in CHD)
  - J Am Soc Echocardiogr 2014;27

- **RV MPI**
  - Circ Cardiovasc Imaging. 2012;5(5)

---

**RV MPI correlates modestly with CMR RV EF**

- NI RV fxn (S’ 14 cm/sec)
- ↓ RV fxn (S’ 7 cm/sec)

- NI RV fxn
- TOF with ↓ RV fxn
Doppler tissue imaging evaluation of right ventricular function at rest and during dobutamine infusion in patients after repair of tetralogy of Fallot

**Table 2** Echocardiographic data

<table>
<thead>
<tr>
<th>Parameter</th>
<th>A. Controls rest</th>
<th>B. TOF patients rest</th>
<th>C. TOF patients peak dobutamine</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sa (cm/s)</td>
<td>13.7 ± 3.1</td>
<td>11.4 ± 2.8*</td>
<td>17.7 ± 4.7***</td>
</tr>
<tr>
<td>Ea (cm/s)</td>
<td>16.3 ± 3.5</td>
<td>11 ± 3.1**</td>
<td>15.6 ± 3.9***</td>
</tr>
<tr>
<td>Aa (cm/s)</td>
<td>9.2 ± 3.2</td>
<td>8.4 ± 2</td>
<td>14.8 ± 5***</td>
</tr>
<tr>
<td>Sa/Q–Sa (cm/s²)</td>
<td>92.3 ± 29.4</td>
<td>68.8 ± 26.4**</td>
<td>176.8 ± 84.5***</td>
</tr>
<tr>
<td>Mean dP/dt (mmHg/s)</td>
<td>180 ± 74</td>
<td>11.9 ± 5.6***</td>
<td></td>
</tr>
<tr>
<td>RVSVi (L/min/m²)</td>
<td>7.8 ± 3.9</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

25 patients
11±6 (6–27) yo
Dobutamine

Exercise Echocardiography Demonstrates Biventricular Systolic Dysfunction and Reveals Decreased Left Ventricular Contractile Reserve in Children After Tetralogy of Fallot Repair

S. Lucy Roche, MB ChB, MRCPCH, MD, Lars Grosse-Wortmann, MD, Mark K. Friedberg, MD, Andrew N. Redington, MD, FRCP, Derek Stephens, MSC, and Paul F. Kantor, MBBCh, DCH, FRCP, Toronto, Ontario, and Edmonton, Alberta, Canada

<table>
<thead>
<tr>
<th>Variable</th>
<th>Controls (n = 27)</th>
<th>Patients with TOF (n = 29)</th>
<th>P</th>
<th>Percentage of patients with TOF with measurements ≥ 2 SDs below controls’ mean</th>
</tr>
</thead>
<tbody>
<tr>
<td>Peak heart rate</td>
<td>164 ± 15.1</td>
<td>137.2 ± 15.6</td>
<td>.0005</td>
<td>41</td>
</tr>
<tr>
<td>RVOT FS (%)</td>
<td>37.7 ± 7.8</td>
<td>25.3 ± 7.1</td>
<td>&lt;.0001</td>
<td>32</td>
</tr>
<tr>
<td>RV LAX FS (%)</td>
<td>34.6 ± 7.4</td>
<td>21.2 ± 6.1</td>
<td>&lt;.0001</td>
<td>48</td>
</tr>
<tr>
<td>RV IVA (m/sec²)</td>
<td>10.3 ± 2.0</td>
<td>3.5 ± 1.4</td>
<td>&lt;.0001</td>
<td>100</td>
</tr>
<tr>
<td>RV Sm (cm/sec)</td>
<td>13.1 ± 2.5</td>
<td>9.2 ± 2.5</td>
<td>&lt;.0001</td>
<td>34</td>
</tr>
<tr>
<td>LV LAX FS (%)</td>
<td>22.3 ± 4.6</td>
<td>23.0 ± 6.3</td>
<td>NS</td>
<td>7</td>
</tr>
<tr>
<td>LV IVA (m/sec²)</td>
<td>9.9 ± 2.6</td>
<td>3.8 ± 1.6</td>
<td>&lt;.0001</td>
<td>75</td>
</tr>
<tr>
<td>LV Sm (cm/sec)</td>
<td>9.9 ± 1.7</td>
<td>8.1 ± 2.4</td>
<td>.001</td>
<td>25</td>
</tr>
</tbody>
</table>

Figure 3 LV force-frequency relationships in asymptomatic children with TOF and healthy controls. Each light blue line represents an individual study participant, and the group locally weighted scatterplot smoothing average curves are superimposed as heavier lines.

Roche et al. J Am Soc Echocardiogr 2015;28:294-301
CMR in CHD

- Anatomical delineation
- Static and Cine imaging
- High–spatial resolution 3D reconstruction
- Phase-contrast (PC) for blood flow measurements
- Late gadolinium enhancement
- Accurate, objective, reproducible and quantitative measurements
  - biventricular size and function
  - pulmonary and systemic blood flow measurements
  - differential PA flow
  - valve regurgitant volumes (e.g., PR, AR)
  - myocardial viability and presence of scarring
  - origin & proximal course of coronaries
**39.55 ml**

**37.8 ml**

**68.69 ml**

**68.5 ml**

**36.9 ml**
Gadolinium uptake in areas of fibrosis 10 to 20 min after contrast injection
Criteria for PVR - Role of CMR

- Asymptomatic patients with ≥ 2 of:
  1. RVEDV index > 150 ml/m² or Z-score > 4 (large patients RV/LV EDV > 2)
  2. RV end-systolic volume index > 80 ml/m²
  3. RV EF < 47%
  4. LV EF < 55%
  e. Large RVOT aneurysm
  5. QRS duration > 140 ms
  6. Sustained tachyarrhythmia related to right heart volume load
  7. Other:
     - RVOT obstruction with RVp ≥ 2/3 systemic
     - Branch PA stenosis (<30% flow to affected lung) not amenable to stenting
     - ≥ Moderate TR
     - Residual ASD or VSD with Qp/Qs ≥ 1.5
     - Severe aortic regurgitation
     - Severe aortic dilatation (diameter ≥ 5 cm)

- Symptomatic patients (exercise intolerance or heart failure) and ≥ 1 of the above
TOF - Cardiac CT

- Anatomical delineation, excellent spatial resolution
- Static and Cine imaging
- 3D reconstruction
- ↑ delineation of small vessels (coronaries, distal PA branches)
- Compatible with pacemakers and defibrillators
- Less artifacts by stainless-steel metallic artifacts
- Radiation, No hemodynamic information
TOF Nuclear Scintigraphy

- Ventricular size & function
- Pulmonary perfusion
- Quantification of cardiac shunts
- Quantification of differential PA blood flow
- V-scan to assess ventilation-perfusion mismatch
- Myocardial perfusion and viability
- Radiation

- Only used in contraindications to CMR and cardiac CT
TOF X-ray Angiography

- Imaging RVOT/PAs/Aorta
- RV size & function / PI qualitatively
- Hemodynamics
- Coronaries (adults, abnormal course, Melody implantation)
- Catheter interventions:
  - PA balloon dilation and stenting
  - percutaneous PV implantation (Melody/Sapien)
  - occlusion of aortopulmonary collaterals
  - closure of residual septal defects
  - coronary artery interventions
- Critically ill patient
- Invasive, radiation, cost
Repaired TOF – Angiography
Systemic RV - CCTGA
CCTGA – Adult algorithm

- Adult adolescent CCTGA patient with systemic RV
  - High degree heart block
    - Biventricular pacing
      - Permanent pacemaker
  - Progressive SRV dysfunction
    - Pharmacological management (reduction of pre- and afterload, inotropic support)
      - Significant LV decompression, shift of the IVS towards the LV
        - PA banding (age dependent)
        - Regular followup (Echocardiography, MRI, treadmill test)
      - No signs of LV decompression
        - Regular followup
  - Progressive TR
    - Preserved SRV function (EF>40%)
      - PA banding (if necessary, age dependent)
    - Decreased SRV function (SRVEF<40%)
      - Early TV replacement
        - Late TV replacement, high risk of RVF
          - CRT (if needed)
  - Atrial/ supraventricular tachyarrhythmias
    - Pharmacological management, Holter monitoring
      - CRT, if needed (QRS >120ms)
      - Implantation of a VAD
        - Bridge to Heart Transplantation
  - Regular followup
Situs inversus and CCTGA: Increased global myocardial thickening and base-to-apex shortening with significant decrease in RV end-systolic volume

Transposition of the Great Arteries

A Atrial Switch

B Arterial Switch

Rastelli
27 patients

Age 29±7 yo

Dobutamine

Positive correlation of rest RV free wall long-axis excursion at rest with CPET

Univentricular Heart - Fontan

- Passive flow of blood through connection of SVC and IVC directly to the pulmonary arteries.
- Use of the normal size ventricle (right or left) as systemic pump.
Dobutamine Stress Echocardiography for the Evaluation of Cardiac Reserve Late After Fontan Operation

10 patients
Age: 28 ±5 yo

Two-Dimensional Global Longitudinal Strain Rate Is a Preload Independent Index of Systemic Right Ventricular Contractility in Hypoplastic Left Heart Syndrome Patients After Fontan Operation

<table>
<thead>
<tr>
<th></th>
<th>Baseline</th>
<th>Dobutamine</th>
<th>Baseline vs Dobutamine</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Steady State</td>
<td>Preload Maneuver</td>
<td>P</td>
</tr>
<tr>
<td>Global strain, %</td>
<td>52</td>
<td>-17.70±3.36</td>
<td>-16.89±3.83</td>
</tr>
<tr>
<td>Global strain rate, s⁻¹</td>
<td>52</td>
<td>-1.30±0.29</td>
<td>-1.34±0.34</td>
</tr>
<tr>
<td>Regional strain, %</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Basal septal</td>
<td>50</td>
<td>-12.5±6</td>
<td>-12.7±6.5</td>
</tr>
<tr>
<td>Mid septal</td>
<td>50</td>
<td>-17.6±4.8</td>
<td>-17.1±5.4</td>
</tr>
<tr>
<td>Apical septal</td>
<td>39</td>
<td>-21.1±8.4</td>
<td>-19.1±8</td>
</tr>
<tr>
<td>Apical lateral</td>
<td>45</td>
<td>-18.6±7.7</td>
<td>-17.2±8.4</td>
</tr>
<tr>
<td>Mid lateral</td>
<td>50</td>
<td>-19.0±5.2</td>
<td>-17.8±5.8</td>
</tr>
<tr>
<td>Basal lateral</td>
<td>49</td>
<td>-18.7±5.8</td>
<td>-18.9±5.2</td>
</tr>
<tr>
<td>Regional strain rate, s⁻¹</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Basal septal</td>
<td>51</td>
<td>-1.12±0.51</td>
<td>-1.23±0.74</td>
</tr>
<tr>
<td>Mid septal</td>
<td>50</td>
<td>-1.31±0.39</td>
<td>-1.32±0.39</td>
</tr>
<tr>
<td>Apical septal</td>
<td>39</td>
<td>-1.55±0.61</td>
<td>-1.61±0.72</td>
</tr>
<tr>
<td>Apical lateral</td>
<td>45</td>
<td>-1.3±0.44</td>
<td>-1.38±0.57</td>
</tr>
<tr>
<td>Mid lateral</td>
<td>50</td>
<td>-1.25±0.36</td>
<td>-1.26±0.41</td>
</tr>
<tr>
<td>Basal lateral</td>
<td>49</td>
<td>-1.3±0.37</td>
<td>-1.29±0.38</td>
</tr>
</tbody>
</table>

52 patients
Median age: 6.6 (2.9–22) yrs

## Surveillance frequency in repaired CHD

### Multimodality Imaging Guidelines for Patients with Repaired Tetralogy of Fallot: A Report from the American Society of Echocardiography Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society for Pediatric Radiology

<table>
<thead>
<tr>
<th>Modality</th>
<th>Age (y)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>&lt;2 y*</td>
</tr>
<tr>
<td></td>
<td>2–9</td>
</tr>
<tr>
<td></td>
<td>10–19</td>
</tr>
<tr>
<td></td>
<td>20–49</td>
</tr>
<tr>
<td></td>
<td>≥50</td>
</tr>
<tr>
<td>Echocardiography</td>
<td>12 mo</td>
</tr>
<tr>
<td>CMR</td>
<td>Not recommended routinely; ordered to address specific questions not answered by echocardiography</td>
</tr>
<tr>
<td></td>
<td>• 36 mo in stable patients</td>
</tr>
<tr>
<td></td>
<td>• 12 mo if moderate (≥150 mL/m²) or progressive (increase of &gt;25 mL/m²) RV dilatation or dysfunction (RV EF ≤ 48% or ≥6% decrease in EF)</td>
</tr>
<tr>
<td>CT</td>
<td>Not recommended routinely; ordered when CMR is indicated but cannot be performed (e.g., metallic artifacts or contraindications to CMR)</td>
</tr>
<tr>
<td>Lung perfusion scan</td>
<td>If predicted RV systolic pressure 60% systemic or smallest branch PA diameter Z score &lt; −2.5; in patients ≥ 10 y of age, consider CMR flow measurements</td>
</tr>
<tr>
<td>X-ray angiography</td>
<td>Not recommended routinely†; ordered when noninvasive methods either cannot be performed or have failed to provide satisfactory diagnostic data</td>
</tr>
<tr>
<td>Chest radiography</td>
<td>Not recommended routinely; may be ordered for evaluation of stent integrity</td>
</tr>
</tbody>
</table>

* Coronary angiography when clinically indicated

*† Repositioning of the right heart catheter may be necessary for adequate CMR imaging.

---

J Am Soc Echocardiogr 2014;27:111-41
## Comparison of Imaging Modalities

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Echocardiography</th>
<th>CMR</th>
<th>CT</th>
<th>Nuclear scintigraphy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Availability</td>
<td>+++</td>
<td>++</td>
<td>++</td>
<td>+++</td>
</tr>
<tr>
<td>Portability</td>
<td>+++</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Cost (relative value units)*</td>
<td>9.11↑</td>
<td>22.51↑</td>
<td>14.39↑</td>
<td>13.59↑</td>
</tr>
<tr>
<td>Radiation risk</td>
<td>-</td>
<td>-</td>
<td>+++</td>
<td>+++</td>
</tr>
<tr>
<td>Artifacts from stainless-steel implants</td>
<td>+</td>
<td>+++</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Sedation requirements in young children</td>
<td>++</td>
<td>+++</td>
<td>+++</td>
<td>++</td>
</tr>
<tr>
<td>Spatial resolution (mm)</td>
<td>&lt;1</td>
<td>1-2</td>
<td>&lt;1</td>
<td>5-10</td>
</tr>
<tr>
<td>Temporal resolution (msec)</td>
<td>20</td>
<td>30</td>
<td>75-175</td>
<td>-</td>
</tr>
<tr>
<td>RV size/function</td>
<td>++</td>
<td>+++</td>
<td>+++</td>
<td>+</td>
</tr>
<tr>
<td>RV pressure</td>
<td>+++</td>
<td>++</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>TR severity</td>
<td>+++</td>
<td>+++</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Mechanism of TR</td>
<td>++++</td>
<td>++</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>PR severity</td>
<td>++</td>
<td>+++</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Branch PAs flow quantification</td>
<td>-</td>
<td>+++</td>
<td>-</td>
<td>+++</td>
</tr>
<tr>
<td>LV size/function</td>
<td>+++</td>
<td>+++</td>
<td>+++</td>
<td>++</td>
</tr>
<tr>
<td>Coronary origins and proximal course</td>
<td>++</td>
<td>+++</td>
<td>+++</td>
<td>-</td>
</tr>
<tr>
<td>Aortic dimensions</td>
<td>+++</td>
<td>+++</td>
<td>+++</td>
<td>+</td>
</tr>
<tr>
<td>Residual shunts</td>
<td>+++</td>
<td>+++</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Pulmonary-to-systemic flow ratio</td>
<td>+</td>
<td>+++</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Aortopulmonary collateral vessels</td>
<td>-</td>
<td>+++</td>
<td>+++</td>
<td>-</td>
</tr>
<tr>
<td>Myocardial viability</td>
<td>+</td>
<td>+++</td>
<td>-</td>
<td>+++</td>
</tr>
</tbody>
</table>

* The cost values are in thousands of dollars.
Conclusions

- Multimodality approach in imaging in repaired CHD
- No single test gives all necessary information
- Choice influenced by:
  - age, need for sedation, acoustic windows
  - clinical question
  - local availability and expertise
  - cost
  - radiation exposure
  - pacemakers/defibrillators
- Research to identify predictors of deterioration vs stable course in this growing population
TOF CMR

- Steady-state free precession (SSFP) →, which is a type of gradient-echo technique characterized by high signal-to-noise ratio, high T2/T1 contrast ratio, and sharp borders between the blood pool and the myocardium.

- Electrocardiographically gated SSFP can be used as a cine magnetic resonance sequence, which is typically used for assessment of ventricular size and function, valve function, and intracardiac and extracardiac anatomy.

- Electrocardiographically gated, respiratory-navigated SSFP sequence can yield a high–spatial resolution static 3D data set, which is often used for detailed assessment of intracardiac anatomy and/or coronary artery anatomy.

- Electrocardiographically gated turbo (fast) spin-echo (TSE) imaging offers high spatial resolution (submillimeter in-plane), excellent contrast between elements of soft tissue, and decreased sensitivity to metallic artifacts compared with gradient-echo sequences, although it provides only static images.

- Contrast-enhanced magnetic resonance angiography (MRA) represents a robust 3D technique.

- Electrocardiographically gated, phase-contrast (PC) flow measurements are used for measurements of blood flow, including flow rates in the great arteries and veins, differential PA flow, and regurgitant volumes (e.g., PR, AR).
CMR - Myocardial Enhancement

Myocardial disarray and plexiform fibrosis
Myocardial disarray and plexiform fibrosis
Comprehensive ECHO views

- Subxiphoid coronal
- 4 chamber
- Parasternal short axis
- Suprasternal short axis
- Subxiphoid sagittal
- Parasternal long axis
- Suprasternal long axis
- Suprasternal right

Υποξιφοειδική Απεικόνιση - Στεφανιαίες τομές
Υποξιφοειδική λήψη - Οβελιαίες τομές
Παραστερνική Τομή κατά το Μακρύ Άξονα
Παραστερνική Λήψη κατά το Βραχύ Άξονα
Κορυφαία Λήψη Τεσσάρων Κοιλοτήτων
Στεφανιαία Υπερστερνική Λήψη
Οβελιαία Υπερστερνική Λήψη
Ventricular Septal Defects (VSD)
Ventricular Septal Defects (VSD)

- Perimembranous: 80%
- Supracristal or conal (outlet): 5-7%
- Muscular: 5-20%
- Trabecular: Marginal (swiss cheese)
- Inlet: 8%
- Apical
GUIDELINES AND STANDARDS

Multimodality Imaging Guidelines for Patients with Repaired Tetralogy of Fallot: A Report from the American Society of Echocardiography Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society for Pediatric Radiology

Anne Marie Valente, MD, FASE, Co-Chair, Stephen Cook, MD, Pierluigi Festa, MD, H. Helen Ko, BS, RDMS, RDCS, FASE, Rajesh Krishnamurthy, MD, Andrew M. Taylor, MD, Carole A. Warnes, MD, Jacqueline Kreutzer, MD, and Tal Geva, MD, FASE, Co-Chair, Boston, Massachusetts; Pittsburgh, Pennsylvania; Massa, Italy; New York, New York; Houston, Texas, London, United Kingdom; Rochester, Minnesota

(J Am Soc Echocardiogr 2014;27:111-41.)
# Imaging surveillance frequency in repaired TOF

<table>
<thead>
<tr>
<th>Modality</th>
<th>&lt;2 y*</th>
<th>2-9</th>
<th>10-19</th>
<th>20-49</th>
<th>≥50</th>
</tr>
</thead>
<tbody>
<tr>
<td>Echocardiography</td>
<td>12 mo</td>
<td>12 mo</td>
<td>24 mo</td>
<td>24 mo</td>
<td>24 mo</td>
</tr>
<tr>
<td>CMR</td>
<td>Not recommended routinely; ordered to address specific questions not answered by echocardiography</td>
<td></td>
<td>36 mo in stable patients</td>
<td>12 mo if moderate (≥150 mL/m²) or progressive (increase of &gt;25 mL/m²) RV dilatation or dysfunction (RV EF ≤ 48% or ≥6% decrease in EF)</td>
<td></td>
</tr>
<tr>
<td>CT</td>
<td>Not recommended routinely; ordered when CMR is indicated but cannot be performed (e.g., metallic artifacts or contraindications to CMR)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lung perfusion scan</td>
<td>If predicted RV systolic pressure 60% systemic or smallest branch PA diameter Z score &lt; −2.5; in patients ≥10 y of age, consider CMR flow measurements</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>X-ray angiography</td>
<td>Not recommended routinely†; ordered when noninvasive methods either cannot be performed or have failed to provide satisfactory diagnostic data</td>
<td></td>
<td></td>
<td>Coronary angiography when clinically indicated</td>
<td></td>
</tr>
<tr>
<td>Chest radiography</td>
<td>Not recommended routinely; may be ordered for evaluation of stent integrity</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Goals of Imaging

- Intracardiac and extracardiac shunts
- TR (degree and mechanism), estimated RVp
- RV evaluation - size and function
  - regional RV wall motion abnormalities
  - RVOT (obstruction and/or aneurysm)
- Degree of PR
- Assessment of the main and branch PAs
- LV size and function
- Aorta - size of the aortic root and ascending aorta
  - degree of AR
  - aortic arch sidedness
- Origin and proximal course of LCA/RCA
- Systemic-to-pulmonary collateral vessels
- Assessment of myocardial viability
Tetralogy of Fallot

= anterior malalignment of the outlet septum

1. Ventricular septal defect
2. Narrow RV outflow tract
3. Aorta overriding interventricular septum
4. Right ventricular hypertrophy
Conduit stenosis and RV fxn -

Exercise Stress Echocardiographic Assessment of Outflow Tract and Ventricular Function in Patients With an Obstructed Right Ventricular-to-Pulmonary Artery Conduit After Repair of Conotruncal Heart Defects

• 35 patients with conduits
• Median age 17 (6-56) yo
• - 25 Fallot
  - 9 truncus arteriosus
  - 6 TGA

TOF – X-Ray Angiography
TOF - 3D printing

Unrepaired TOF (from ECHO)  Repaired TOF (from CMR)

May be useful in organizing interventions, PV implantation, construction of implantable materials that fit in the patient’s RVOT
• Boot-shaped heart (not common)
• ↓ pulmonary markings in ↓ pulmonary blood flow
CMR – Angio – RVOT / PAs
CMR – Anatomy – Cine – PS/PI
CMR – Cine – RVOT / Flows
CMR – Anatomy - Cine – Tagging
ECHO: 3D / TEE / Stress

- **3D ECHO:**
  - anatomy
  - RV & LV size/fxn (underestimate vs CMR)

- **TEE:**
  - PFO/ASD
  - TTE challenging
  - infective endocarditis
  - guide to interventional procedures

- **Stress ECHO:**
  * Stress Echo 2020 study with TOF branch (recruits since 2016)
    - assess RV contractile reserve
    - correlation with severity (NYHA, BNP, peak VO₂, 6MWD)
    - assess medium and long-term prognostic value of SE
Tetralogy of Fallot

Usefulness of Dobutamine Stress Echocardiography with Tissue Doppler Imaging for the Evaluation and Follow-Up of Patients with Repaired Tetralogy of Fallot

21 patients (median age 27.7 (19-48) years, Dobutamine

Figure 2 (A), Correlation between the baseline values of TDI velocity of the tricuspid annulus during systole (Sa) and the values of Sa at peak infusion rate. (B), Correlation between the baseline values of TDI velocity of the tricuspid annulus during systole (Sa) and the absolute dobutamine-induced increase of Sa (ΔSa).

16 patients (25.6±3.7yo)

- Dobutamine

- At rest, all patients had reduced:
  - IVA
  - s-velocities
  - e-velocities

compared with normal subjects with systemic LV
Pulmonary Hypertension – Master 2 step

Simple stress echocardiography unmask early pulmonary vascular disease in adult congenital heart disease

Annelieke C.M.J. van Riel \textsuperscript{ab}, Rianne H.A.C.M. de Bruin-Bon \textsuperscript{a}, Emma C. Gertsen \textsuperscript{a}, Ilja M. Blok \textsuperscript{ab}, Barbara J.M. Mulder \textsuperscript{ab}, Berto J. Bouma \textsuperscript{a,*}

\textsuperscript{a} Department of Cardiology, Academic Medical Centre, Amsterdam, The Netherlands
\textsuperscript{b} ICN – Netherlands Heart Institute, Utrecht, The Netherlands

- 78 patients (43.2 ± 14.5 yo)
- Symptom-limited Master two-step test
- Systolic PAP: TR CW + RAP (from IVC size and collapsibility)
- Mean PAP = 0.6 \times \text{systolic PAP} + 2
- Suspected early PVD: mean PAP > 34 mmHg
  - 16 patients (21%)
  - CO > 10 L/min
  - mean PAP/CO > 3

Van Riel et al. IJC 197 (2015) 312–314
Pulmonary Hypertension - Bike

Right ventricular load and function during exercise in patients with open and closed atrial septal defect type secundum

- 20 open ASDs
  39.3±17.5 yo
- 30 closed ASDs
  42.4±16.8 yo

Figure 4. If right ventricular RVFAC increases ≤5% from rest to peak exercise, open and closed atrial septal defect (ASD) patients present with a significantly higher mean pulmonary artery pressure (mPAP) at peak exercise.
Conclusions

- Indications for Stress ECHO in pediatric and CHD are evolving
- Serial testing may provide help in:
  - diagnosis
  - risk stratification
  - follow-up
  - evaluation of treatment
- The impact of SE on patient outcomes is under research
- Efficiency may be increased with comprehensive TTE
- Deformation and multidimensional imaging may ↑ utility of SE
- SE has great versatility and its use is likely to expand