Assessing the Patient Following Repair for Congenital Disease

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No relevant disclosures
## What to look for after repair: Simple defects

<table>
<thead>
<tr>
<th>Defect</th>
<th>Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>VSD</td>
<td>Residual shunt around a patch (endocarditis risk)</td>
</tr>
<tr>
<td></td>
<td>Alternative or additional diagnoses</td>
</tr>
<tr>
<td>AVSD</td>
<td>Mitral stenosis or regurgitation</td>
</tr>
<tr>
<td></td>
<td>LVOT stenosis</td>
</tr>
<tr>
<td></td>
<td>Small residual shunts</td>
</tr>
<tr>
<td>Pulm Stenosis</td>
<td>Pulmonary valve regurgitation (post valvotomy)</td>
</tr>
<tr>
<td>Coarctation</td>
<td>Recoarctation, Pseudoaneurysm</td>
</tr>
<tr>
<td>Ebstein repair</td>
<td>Valve dysfunction, RV systolic dysfunction</td>
</tr>
</tbody>
</table>
Assessing Repaired Complex Defects

- Tetralogy
- Transposition
- Fontan
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

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111
Tetralogy of Fallot

Parasternal Long Axis

Normal
Tetralogy of Fallot

Parasternal Long Axis

Normal

Tetralogy/Pulm Atresia/Truncus
Tetralogy of Fallot

RV hypertrophy, septal bounce in early diastole
Tetralogy of Fallot

Pulmonary valve stenosis

PV Vmax = 1.79 m/sec
Pk Grad = 12.9 mmHg
Tetralogy of Fallot

Spectral Doppler

Severe PR

PV Vmax = 1.79 m/sec
Pk Grad = 12.9 mmHg
Tetralogy of Fallot

Severe pulmonary valve regurgitation
Tetralogy of Fallot

Assessment of RV size
Tetralogy of Fallot

Assessment of RV size
Tetralogy of Fallot

Assessment of RV size
Tetralogy of Fallot

Severe RV enlargement
Tetralogy of Fallot

Severe RV enlargement
Tetralogy of Fallot

RV enlargement: RVOT view
Estimating RV volume

Rudski LG, J Am Soc Echocardiogr 2010;23:685-713
RV Volume Methodology

ASE RV guidelines were shown to be less reliable in congenital patients

Volume accuracy improves with 3D techniques
van der Zwaan HB, Eur J Echocardiogr. 2011 Sep;12(9):656-64

3d still underestimates volume relative to MRI
Crean AM, J Cardiovasc Magn Reson. 2011 Dec 8;13:78

Use All Available Information
Tetralogy of Fallot

What not to miss

- Severe PR
- RV enlargement (not just the 4 chamber view)
- LV dysfunction
Semilunar Valve Orientation

Anterior aorta, parallel valve planes
Types of Transposition

L-TGA

D-TGA
Types of Transposition

L-TGA

D-TGA

Atrial switch

Arterial Switch

Before ~1980

After ~1990
Types of Transposition

L-TGA

D-TGA

Atrial switch

Arterial Switch

Before 1980

After 1990
# Ventricular Morphology

<table>
<thead>
<tr>
<th>Morphologic RV</th>
<th>Morphologic LV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heavily trabeculated</td>
<td>Smooth walled (compacted)</td>
</tr>
<tr>
<td>Moderator band</td>
<td>Fewer trabeculations</td>
</tr>
<tr>
<td>Apical AV valve plane</td>
<td>Basal AV valve plane</td>
</tr>
</tbody>
</table>

Useful to differentiate
atrial from arterial switch
Assessment of Systemic Right Ventricle

L-TGA or D-TGA with Atrial Switch
RV functional assessment

Non-geometric methods
  TAPSE
  Fractional area change
dP/dt
TDI
MPI
Strain
RV functional assessment

Non-geometric methods

TAPSE  Most precise/reproducible
Fractional area change
\( \text{dP/dt} \)
TDI
MPI
Strain

Iriart X, Arch Cardiovasc Dis. 2012 Aug-Sep;105(8-9):432-41
RV functional assessment

Non-geometric methods

- TAPSE
- Fractional area change
- $dP/dt$
- TDI
- MPI
- Strain

Most accurate relative to CMR

Khattab K, Am J Cardiol. 2012 Dec 28 Best performers
RV functional assessment

Non-geometric methods

TAPSE
Fractional area change
dP/dt
TDI
MPI
Strain

Most predictive of clinical events

Kalogeropoulos AP, JASE 2012 Mar;25(3):304-12
Systemic Right Ventricle

Severe TR (systemic AV valve)
Systemic Right Ventricle

Septal displacement

RV  LV
Systemic Right Ventricle

Septal displacement
Systemic Right Ventricle

Septal displacement
Systemic Right Ventricle

Benefits of sub-pulmonary stenosis
D-TGA with Atrial Switch Palliation

Atrial baffle stenosis or leak
Atrial baffle stenosis or leak
D-TGA with Atrial Switch Palliation

Atrial baffle stenosis or leak: Look for bubbles on the systemic side
D-TGA with Arterial Switch
D-TGA with Arterial Switch

Lacompte maneuver: Pulmonary bifurcation anterior to aorta
D-TGA with Arterial Switch

Neoaortic root enlargement
D-TGA with Arterial Switch

Neoaortic root enlargement

Neoaortic valve regurgitation
Transposition

What not to miss

Clarify the anatomy (L-TGA vs. D-TGA)
Clarify the repair (arterial switch vs. atrial switch)

Atrial Switch (Mustard or Senning repair)
  Baffle leak or stenosis
  TR structure and function

Arterial Switch (Jatene/LeCompte)
  Segmental wall motion (coronary artery stenosis)
  Pulmonary stenosis
  Neoaortic valve regurgitation
Tetralogy
Transposition
Fontan
“Single” Ventricle / “Univentricular” Heart
“Single” Ventricle / “Univentricular” Heart
Fontan palliation

Passive venous flow to the pulmonary capillary bed
Fontan palliation

??????????
Fontan palliation

Key Assessments

Mitral or tricuspid valve regurgitation

Ventricular systolic function (usually normal)

Large venous clot or obstruction
Fontan palliation

Classic atrio pulmonary connection
Fontan palliation

Classic atriopulmonary connection
Fontan palliation

Classic atriopulmonary connection: atrial thrombosis
Fontan palliation

Extracardiac conduit
Fontan palliation

Fenestration
Fontan palliation

Normal ventriculoarterial relationship
Fontan palliation

Transposed great arteries, VSD, subaortic stenosis
Single Ventricle/Fontan

What not to miss

Type of ventricular anatomy *(check the history!!)*

Function of the atrioventricular valve(s)

Small fenestration

Visible thrombi
Key Points to Remember

Use the patient’s history to help

Keep common complications in mind

Describe what you see
Thank You