The role of cardiovascular magnetic resonance (CMR) in evaluating the adult population with congenital heart disease continues to expand. This pocket guide aims to provide a day-to-day companion for those new to congenital CMR and for those looking for a quick reference guide in routine practice. The booklet gives an overview of the most common abnormalities and interventions as well as post-operative complications. It also provides typical scan protocols, key issues and a guide for reporting for each topic.

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Ananth Kidambi
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The Cardiovascular Magnetic Resonance Pocket Guide represents the views of the ESC Working Group on Cardiovascular Magnetic Resonance and was arrived at after careful consideration of the available evidence at the time it was written. Health professionals are encouraged to take it fully into account when exercising their clinical judgment. This pocket guide does not, however, override the individual responsibility of health professionals to make appropriate decisions in the circumstances of the individual patients, in consultation with that patient and, where appropriate and necessary, the patient's guardian or carer. It is also the health professional's responsibility to verify the applicable rules and regulations applicable to drugs and devices at the time of prescription.

We acknowledge the support and advice we have received from Emanuela Valsangiacomo Buechel and James Oliver.
Standard Views
- Anatomical Stacks -

Transaxial stack

Sagittal stack

Coronal stack
Standard Views
- Cine Imaging -

Transaxial stack

Transaxial stack

pVLA

pVLA

pSA

pSA

HLA

HLA

VLA

Simplified planning:
Use pVLA, pHLA and pSA instead of VLA, HLA and SA (see LV stack)

Synonyms:
VLA = 2CH; HLA = 4CH
Standard Views
- Cine Imaging -

HLA  VLA  SA cine stack

Coronal localizer  Sagittal localizer  Transaxial cine stack

HLA  VLA  Atrial cine stack*

* RV measurements alternatively from SA LV stack
  Important is consistency for reproducibility
  * Alternatively use HLA stack
Standard Views

- Cine Imaging -

Transaxial stack

Sagittal RVOT

Coronal RVOT

Transaxial stack

Proximal LPA°

Distal LPA°

Transaxial stack

Proximal RPA°

Distal RPA°

° Full bifurcation planned from transaxial and coronal localizer
Standard Views
- Cine Imaging -

Basal SA of LV stack  VLA through apex  Sagittal LVOT

Sagittal LVOT  Coronal LVOT

Sagittal LVOT  Coronal LVOT  Aortic valve /root
Standard Views
- Cine Imaging -

Transaxial
Transaxial
Aortic arch

HLA
RVOT
RV in-/outflow

3-point planning

Index
Standard Views
- Flow Imaging -

Sagittal LVOT  Coronal LVOT  Ao flow
Sagittal RVOT  Coronal RVOT  MPA flow
Transaxial stack  LPA cross-cut  LPA flow°
Transaxial stack  RPA cross-cut  RPA flow°

Simplified planning:
° Directly from coronal and axial localizer
Standard Views
- Flow Imaging -

Modified sagittal Basal SA of LV stack ASD flow

Basal SA of LV stack Coronal localizer VSD flow

Coronal localizer Transaxial magnitude Transaxial flow

descAo Azygous Hemi-Azygous
descAo Azygous Hemi-Azygous
## Arrhythmia

<table>
<thead>
<tr>
<th>Technique</th>
<th>Comment</th>
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<tbody>
<tr>
<td>Ensure correct lead position</td>
<td>You might be lucky!</td>
</tr>
<tr>
<td>Try again</td>
<td>Use beta-blockers or other antiarrhythmic medication</td>
</tr>
<tr>
<td><strong>Heart rate</strong> and/or <strong>rhythm control</strong> before scanning</td>
<td>Increases breath-hold time</td>
</tr>
<tr>
<td>Use <strong>arrhythmia rejection</strong></td>
<td>Cardiac diastole is not entirely visualised</td>
</tr>
<tr>
<td>Use <strong>prospective triggering</strong></td>
<td></td>
</tr>
<tr>
<td>Use <strong>real-time imaging</strong></td>
<td>Reduces temporal and spatial resolution as well as SNR</td>
</tr>
<tr>
<td>Scan <strong>in inspiration</strong></td>
<td>If heart signal capture is suboptimal</td>
</tr>
<tr>
<td><strong>Increase NSA</strong></td>
<td>Reduces vasovagal arrhythmias</td>
</tr>
<tr>
<td><strong>Alternative sequence</strong></td>
<td>Signal averaging can be useful for e.g. delayed contrast imaging</td>
</tr>
<tr>
<td></td>
<td>E.g. turbo field echo rather than SSFP, white blood imaging rather than black blood sequences</td>
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</table>
## Poor Breath-Holders

<table>
<thead>
<tr>
<th>Acceleration technique</th>
<th>Comment</th>
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<tbody>
<tr>
<td>• Reduce <strong>number of slices</strong> acquired per breath-hold</td>
<td>• Increases overall scan time</td>
</tr>
<tr>
<td>• Reduce <strong>number of phases</strong> for each breath-hold:</td>
<td>• Reduces SNR</td>
</tr>
<tr>
<td>- by reducing <strong>acquisition matrix</strong> (scan or phase percentage)</td>
<td>• Increases spatial resolution</td>
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<td>- by reducing <strong>FOV</strong></td>
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<tr>
<td>• Increase <strong>voxel size</strong></td>
<td>• Decreases spatial resolution</td>
</tr>
<tr>
<td>• Use <strong>parallel imaging</strong></td>
<td>• Prone to artefacts</td>
</tr>
<tr>
<td>• Use <strong>respiratory navigator</strong></td>
<td>• Increases overall scan time</td>
</tr>
<tr>
<td>• Acquire images in <strong>inspiration</strong></td>
<td>• Varying slice position with each breath-hold</td>
</tr>
<tr>
<td>• Use <strong>real time imaging</strong></td>
<td>• Reduces image quality</td>
</tr>
<tr>
<td>• Consider <strong>general anaesthesia</strong></td>
<td></td>
</tr>
<tr>
<td>• Ensure <strong>correct understanding</strong> of breath-hold technique</td>
<td>• If patient has no respiratory problems</td>
</tr>
<tr>
<td>• Ensure correct understanding of breath-hold technique</td>
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</tbody>
</table>
Sequential Segmental Analysis
- Overview -

• Important to start report with comprehensive segmental analysis

Cardiac Situs
- Situs solitus
- Situs inversus
- Situs ambiguous
  Based on atrial morphology
  Bronchial situs is a surrogate

Cardiac Position
- Levocardia
- Dextrocardia
- Mesocardia
  Based on position in thorax

Cardiac Segments
- Atrial Segment
- Ventricular Segment
- Arterial Segment

Connections
- Veno-Atrial Connection
- Atrio-Ventricular Connection
- Ventricular-Arterial Connection

Taken together, segmental analysis can describe any congenital heart disease
Sequential Segmental Analysis
- Cardiac Situs -

Cardiac Situs

- **Situs Solitus**
  - Anterior RA (on right)
  - Posterior LA (on left)

- **Situs Inversus**
  - Posterior RA (on left)
  - Anterior LA (on right)

- **Situs Ambiguous**
  - Right
  - Left
  - RA or LA isomerism

Abdominal Situs

- **Situs Solitus**
- **Situs Inversus**
- **Heterotaxia**
Sequential Segmental Analysis
- Cardiac Position -

Cardiac Position

Dextroposition  Mesoposition  Levoposition

Cardiac Orientation

Dextrocardia  Mesocardia  Levocardia
### Atrial Segment

- **Right atrium**
  - Broad, based, triangular appendage
  - Short and vertical bronchus arrangement

- **Left atrium**
  - Narrow, tubular appendage
  - Long and horizontal bronchus arrangement

### Ventricular Segment

- **Right ventricle**
  - Trabeculated
  - TV associated, TV attachments to the septal moderator band
  - Muscular infundibulum between inlet and outlet

- **Left ventricle**
  - Smooth walled
  - MV associated, MV attachments to papillary muscles

### Arterial Segment

- **Pulmonary Trunk**
  - Bifurcation to RPA and LPA

- **Aorta**
  - Left- or right-sided
  - Coronary arteries
  - Regular branches
Veno-Atrial Connection

- IVC and SVC connections
- Presence of left SVC (90% left SVCs drain to RA via coronary sinus)
- Normal, partial or total anomalous pulmonary venous drainage

Atrio-Ventricular Connection and Inlets

**AV concordance**
- RA is connected to RV, LA to LV

**AV discordance**
- RA is connected to LV, LA to RV

The valves go with the ventricles

Double inlet, mitral atresia, tricuspid atresia, AVSD

Ventricular-Arterial Connection

**VA concordance**
- LV is connected to Ao
- RV to MPA

**VA discordance**
- LV is connected to MPA
- RV to Ao

Double outlet, single outlet

(e.g. pulmonary atresia or truncus arteriosus)
Atrial Septal Defect

**Pre-operative findings**
- ASD
- RA and RV dilation ± RV dysfunction

**Associated findings**
- Partial anomalous pulmonary venous drainage
  - look for right upper pulmonary vein (most common anomaly)
- Mitral valve abnormalities and regurgitation (ostium primum)
- Tricuspid regurgitation

Superior sinus venosus defect
Ostium secundum defect
Ostium primum defect
Inferior sinus venosus defect
Atrial Septal Defect

Interventions

• Percutaneous device (secundum)
• Surgical atrial septal closure

Post-operative complications

• Residual ASD

Protocol

1. Anatomical stack
2. VLA, HLA, LV stack, RV stack
3. LVOT, RVOT
4. MPA flow
5. Ao flow
6. ASD flow
7. MRA pulmonary veins
8. 3D whole heart

Report

1. **Size** (corrected for BSA) and **function**
   • LV: EDV, ESV, SV, EF, RWMA
   • RV: EDV, ESV, SV, EF, RWMA, note any RV dilation
2. **ASD type, size and location**
3. **Qp (MPA flow) : Qs (Ao Flow)**
4. **Pulmonary venous connection**
5. **Associated findings**
Atrial Septal Defect

Key issues

1. ‘Red flags’
   - Significant RV dilatation or dysfunction
   - Qp:Qs > 1.8:1
   - Associated abnormalities e.g. PAPVD
   - Pulmonary hypertension

2. Major types of ASD
   - **Ostium secundum defect**
     - Most common defect, 75% of all ASD cases
     - Fenestrated or netlike septum possible
   - **Ostium primum defect**
     - Atrioventricular septal defect
     - Commonly associated with mitral valve abnormalities, e.g. mitral cleft - common AV valve
   - **Sinus venosus defect**
     - Commonly associated with partial anomalous connection of the right-sided pulmonary veins

3. Calculation of shunting volume
   - SV from MPA flow / SV from Ao flow
   - Alternatively RV SV / LV SV (w/o valve disease)
   - Use MPA /Ao flow and RV SV / LV SV as internal validation
Atrial Septal Defect

Ostium secundum defect
- ASD with left-to-right shunt
- ASD with left-to-right shunt
- ASD jet - VENC

Sinus venosus defect
- Superior SVASD
- Anomalous PV to SVC
- Inferior SVASD

Flow-time curve of ASD with left-to-right shunt: Qp:Qs 2.5:1
Ventricular Septal Defect

Pre-operative findings

• VSD
• LA and LV dilation ± LV dysfunction
• LV and RV dilation with AVSD (shunting at atrial and ventricular levels)

Associated findings

• Peri-membranous VSD: Septal aneurysm, double chambered RV, aortic valve prolapse and aortic incompetence, sub-AS
• AVSD: Common AV valve and regurgitation
# Ventricular Septal Defect

## Interventions
- Surgical closure with ventricular septal patch
- Percutaneous device

## Post-operative complications
- Residual VSD
- RVOT obstruction
- Valvular regurgitation

## Protocol
1. Anatomy stacks
2. VLA, HLA, LV stack, RV stack
3. LVOT, RVOT
4. AoV
5. MPA flow
6. Ao flow
7. VSD flow

## Report
1. **Size** (corrected for BSA) and **function**
   - LV: EDV, ESV, SV, EF, RWMA, note any LV dilation
   - RV: EDV, ESV, SV, EF, RWMA
2. **VSD type, size and location**, VSD jet velocity
3. **Qp (MPA flow) : Qs (Ao Flow)**
4. Associated findings
Ventricular Septal Defect

Key issues

1. ‘Red flags’
   - Non-restrictive
   - Dilated LV ± RV
   - Qp:Qs > 1.8
   - Associated valvular dysfunction

2. Is the VSD restrictive or not?
   - Restrictive VSD
     - Small (<1/2 aortic valve diameter)
     - High left to right velocity
     - Normal RV and pulmonary pressures
   - Non-restrictive
     - Large VSDs
     - RVH and pressure-loaded RV
     - Eisenmenger syndrome if uncorrected

3. If the VSD is small, CMR may not detect it
   - Consider TTE/TEE instead

4. VSD jet
   - Jet size and velocity are dependent on defect size and pressure differences between the chambers
   - Peak velocity often underestimated by CMR
5. **LV or RV dilation?**
   - The VSD shunts the blood directly into the RVOT leading to LV dilation even in large defects
   - + RV dilation only if
     - AVSD
     - Eccentric VSD jet flow through TV into RA
     - End-stage disease with Eisenmenger

6. **Gerbode defect**
   - LV to RA shunt
   - RA ± RV dilation
   - Congenital disorder or after endocarditis or iatrogenic

7. **Calculation of shunting volume**
   - SV from MPA flow / SV from Ao flow
   - Alternatively RV SV / LV SV (in patients w/o valve disease)

8. **Common synonyms**
   - Peri-membranous: infracristal, conoventricular
   - Muscular: trabecular
   - Doubly committed: supracristal, subarterial, outlet
   - AVSD: canal-type, cushion-type, AV-septum type
Ventricular Septal Defect

Muscular VSD

VSD – HLA diastole  VSD jet – HLA systole  VSD jet - SA

Peri-membranous VSD

VSD jet - LVOT  VSD jet – basal SA  Dilated LV – HLA

Doubly committed VSD

VSD jet – basal SA  VSD jet - RVOT  VSD jet – RVOT VENC

Index
Double-Chambered Right Ventricle

Pre-operative findings

- Muscular anomalous sub-pulmonary band dividing the RV cavity into two chambers
- RV hypertrophy (may contribute to sub-pulmonary stenosis)
- VSD (not always present)
  - Often per-membranous
  - Can flow into low or high pressure chamber

Associated findings

- TR (high velocity jet)
- RVOT obstruction
Double-Chambered Right Ventricle

Interventions

• Surgical resection
• VSD closure

Post-operative complications

• Intra-ventricular restenosis

Protocol

1. Anatomy stacks
2. VLA, HLA, LV stack, RV stack
3. LVOT, RVOT
4. RV in- / outflow
5. MPA flow
6. AoV flow
7. Coronal stack
8. VSD flow
9. TR flow

Report

1. Size (corrected for BSA) and function
   - LV: EDV, ESV, SV, EF, RWMA, mass
   - RV: EDV, ESV, SV, EF, RWMA
2. RV muscular band: location (high / low) and severity of stenosis – difficult to assess adequately by CMR
3. LVOT or RVOT obstruction
4. Hypertrophy of proximal / inflow RV chamber
5. VSD type, size, location, Qp (MPA flow):Qs (AoV flow)
6. Presence of TR
Key issues

1. **Anomalous muscle bundle**
   - Usually associated with a VSD; consider spontaneous closure if not present
   - Divides the RV into a prestenotic inflow chamber and a low-pressure infundibular chamber
   - May occur anywhere through RV from adjacent to PV down to apex
   - Best visible on RV in-/outflow or coronal view
   - Can be missed on HLA view

2. **VSD**
   - Most commonly peri-membranous
   - May communicate with either proximal or distal chamber
   - Shunts in the proximal chamber can be underestimated due to the high-pressure status

3. **RVOT obstruction**
   - Due to progressive hypertrophy of RV and muscle bundles
   - May lead to RV failure

4. **TR**
   - High TR jet velocity can be mistaken as PH
Muscular band (arrow) dividing the RV in a high (HP) and a low pressure chamber (LP) - diastole and systole
Pre-operative findings

- PDA
- LA and LV dilation ± LV dysfunction
- Dilated pulmonary veins and ascAo in large PDA

Associated findings

- Occasionally aortic coarctation
## Patent Ductus Arteriosus

### Interventions
- Occluder device
- Coil embolization
- Surgical ligation

### Post-operative complications
- Residual shunt

### Protocol

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<tbody>
<tr>
<td>1</td>
<td>Anatomy stacks</td>
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<tr>
<td>2</td>
<td>VLA, HLA, LV stack, RV stack</td>
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<tr>
<td>3</td>
<td>LVOT, RVOT</td>
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<td>4</td>
<td>PAs</td>
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<td>5</td>
<td>AoArch</td>
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<td>6</td>
<td>PDA cine stack</td>
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<tr>
<td>7</td>
<td>MPA and branch PAs flow</td>
</tr>
<tr>
<td>8</td>
<td>AoV flow</td>
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<tr>
<td>9</td>
<td>Pre-/post PDA aortic flow</td>
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<tr>
<td>10</td>
<td>PA in-plane flow</td>
</tr>
<tr>
<td>11</td>
<td>MRA aorta</td>
</tr>
<tr>
<td>12</td>
<td>3D whole heart</td>
</tr>
</tbody>
</table>

### Report

1. **Size** (corrected for BSA) and **function**
   - LV: EDV, ESV, SV, EF, mass, note any LV dilation
   - RV: EDV, ESV, SV, EF
2. **PDA length, diameter and form** (conical / window / tubular)
3. **Qp (MPA flow) : Qs (AoV flow)**
Patent Ductus Arteriosus

Key issues

1. **Search for PDA if:**
   - Unexplained flow artefacts in PAs or MPA
   - Unexplained LA and LV dilation
   - Continuous machine-like heart murmur
   - Endocarditis with no valvular defects

2. **Calculation of shunting volume**
   - SV from Ao flow / SV from MPA flow
   - Alternatively LV SV / RV SV (in patients w/o valve disease)

3. **Magnitude of the excess pulmonary blood flow** depends on:
   - Diameter and length of PDA
   - Systemic and pulmonary vascular resistance

4. **A large and uncorrected PDA can result in pulmonary hypertension**

5. **Right sided PDA** typically associated with other congenital abnormalities
Patent Ductus Arteriosus

Dilated LV

PDA jet in MPA

Systolic flow of PDA in MPA

Diastolic flow of PDA in MPA
Findings

Classic BT Shunt (CBTS)
- Subclavian artery to PA

Modified BT Shunt (MBTS)
- Gore-Tex tube from subclavian artery to PA
# Blalock-Taussig Shunt

## Late interventions
- BT stent

## Postoperative complications
- BT shunt stenosis
- Aneurysm formation
- PA dilatation
- Pulmonary hypertension if large /excess shunting

## Protocol

<table>
<thead>
<tr>
<th>1.</th>
<th>Anatomy stacks</th>
<th>6.</th>
<th>AoV flow</th>
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<tr>
<td>2.</td>
<td>VLA, HLA, LV stack, RV stack</td>
<td>7.</td>
<td>MPA</td>
</tr>
<tr>
<td>3.</td>
<td>LVOT, RVOT</td>
<td>8.</td>
<td>PAs flow distal to shunt</td>
</tr>
<tr>
<td>4.</td>
<td>PAs</td>
<td>9.</td>
<td>MRA</td>
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<td>5.</td>
<td>Shunt cines and flow</td>
<td>10.</td>
<td>3D whole heart</td>
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</tbody>
</table>

## Report

1. **Size** (corrected for BSA) and **function**
   - LV: EDV, ESV, SV, EF, mass
   - RV: EDV, ESV, SV, EF

2. **BT shunt dimensions, patency** and flow

3. Presence of aneurysm formations

4. Qp (MPA flow) : Qs (AoV flow)
Key issues

1. **Palliative intervention in cyanotic heart disease**
2. May be used as a **bridge** to Glenn / Fontan circulation
3. Shunts may be small, and best seen with MRA (timed to aorta)
4. **Haemodynamic complications**
   - Subclavian steal (vertebrobasilar ischemia)
5. **A number of alternative palliative shunts exist:**
   - Waterston (ascending aorta to RPA)
   - Potts (descending aorta to LPA)
   - Central (aorta to MPA)
   - Cooley (ascending aorta to RPA, intra-pericardial)
Blalock-Taussig Shunt

Classic BT shunt of left subclavian artery to MPA - MRA

Modified BT shunt from the right subclavian artery to MPA - MIP

Bilateral MBT shunt - transaxial

Bilateral MBTS - flow imaging
Aortic Valve Disease

Pre-operative findings

- Aortic valve stenosis and/or regurgitation
- Concentric LV hypertrophy – aortic stenosis
- Dilated LV with excentric hypertrophy – aortic regurgitation
- Dilated LV with impaired systolic function – late stage AS or AR

Associated abnormalities

- Aortic coarctation
- Subaortic- or supravalvular aortic stenosis
- VSD in subaortic stenosis
- Shone Complex: parachute mitral valve, mitral stenosis, BAV, and coarctation of the aorta
Aortic Valve Disease

Interventions

• Aortic valve repair
• Aortic valve replacement (+/- aortic root; ascending aorta)
• **Ross procedure**

Post-operative complications

• Valve dysfunction; Paravalvular regurgitation

Ross procedure

• Aortic insufficiency
• Aortic autograft / ascending aortic dilation
• RVOT obstruction
• Pulmonary allograft stenosis or regurgitation
• Coronary artery stenosis
## Aortic Valve Disease

### Protocol

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<td>AoV, PV</td>
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<td>5.</td>
<td>Aortic arch</td>
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<td>6.</td>
<td>AoV flow</td>
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### Report

1. **Size** (corrected for BSA) and **function**
   - LV: EDV, ESV, SV, EF, mass, note any LV dilation
   - RV: EDV, ESV, SV, EF, RWMA
2. **Aortic stenosis and/or regurgitation**
3. **Aortic dimensions:**
   - LVOT, annulus, SoV, STJ, ascAo, arch, isthmus, descAo
4. **Aneurysm formation**
5. **Post Ross procedure**
   - RV RVOT obstruction, PV stenosis and/or regurgitation, coronary stenosis
6. **Associated pathologies**
Aortic Valve Disease

Key issues

1. CMR has lower spatial and temporal resolution than ultrasound but is a reasonable alternative if poor echo image quality

2. Comprehensive valve assessment:
   • LV / RV dimensions, mass, fibrosis, and function
   • Forward and regurgitant flow / fraction
   • Mean / peak velocity – will underestimate
   • Jet detection, direction and origin
   • Valve area by direct planimetry

3. Ross procedure

   Advantages
   • Longevity of the pulmonary allograft is superior to biological prosthesis
   • Favourable hemodynamics
   • No need for anticoagulation
   • The valve grows as the patient grows

   Disadvantages
   • Single valve disease (aortic) treated with a two valve procedure (aortic and pulmonary)
Aortic Valve Disease

Tricuspid AV

Bicuspid AV in systole
Bicuspid AV in diastole

Quadricuspid AV

Ross procedure - Dilated ascending aorta
Ross procedure – RV homograft stenosis

Ross procedure – proximal autograft anastomosis dehiscence
Ross procedure - Dilated SoV and ascending aorta, AR
Aortic Coarctation

Pre-operative findings
- Narrowing in the region of the ligamentum arteriosum, the arch or the isthmus
- Collaterals
- +/- hypertrophic LV

Associated abnormalities
- Bicuspid AV and dilated ascending aorta
- Subaortic stenosis
- Arch hypoplasia
- VSD
- Mitral valve abnormalities, such as parachute MS
Aortic Coarctation

Post-operative complications

- Restenosis
- Aneurysm formation
- Collaterals

Initial interventions

A) Stent
B) End-to-end anastomosis
C) Interposition graft
D) Patch augmentation
E) Bypass graft
F) Subclavian flap repair
Aortic Coarctation

Protocol

1. Anatomy stack
2. VLA, HLA, LV stack
3. LVOT
4. AoV
5. Aortic arch
6. AoV flow
7. Pre-stenotic flow
8. Post-stenotic peak flow
9. DescAo flow (diaphragm)
10. MRA aorta

Report

1. Size (corrected for BSA) and function
   - LV: EDV, ESV, SV, EF, mass
2. Aortic dimensions:
   - LVOT, annulus, SoV, STJ, ascAo, arch, isthmus, descAo
   - Add cardiac phase, orientation and sequence
3. Severity of stenosis
   - Minimal dimensions
   - Post-stenotic peak flow
   - Presence and degree of collateral flow
4. Aneurysm formation
5. Associated pathologies
Aortic Coarctation

Caveats of aortic measurements

Transaxial
Overestimation due to non-orthogonal plane

Oblique sagittal
Underestimation due to non-central or non-perpendicular plane

Black Blood
Overestimation possible due to inclusion of aortic wall

MRA
Over- / underestimation due to:
• Acquisition not cardiac cycle specific
• Motion artefacts, particular at aortic root /ascAo

3D whole heart
Over- / underestimation due to:
• Lower spatial resolution
• Motion artefacts
Aortic Coarctation

Key issues

1. **Aortic dimensions:**
   - Be aware of caveats of aortic measurements – see above
   - Diastolic measurements from cine images are preferred
   - Be clear in your report, which cardiac phase, orientation and sequence you used for measurements

2. **Severity of coarctation:**
   - Peak systolic flow is often underestimated by CMR - echocardiography superior to CMR
   - Diastolic prolongation of forward flow is a sign of significant coarctation

3. **Collateral flow:**
   - A decrease of <10% (prestenotic - descAo flow) is expected physiologically
   - An increase implies collateral flow rejoining the descending thoracic aorta
   - Abundant collaterals may reduce the gradient across the coarctation and mask the severity of the obstruction

4. **Aneurysms** of the circle of Willis or other cerebral vessels occur in up to 10% of patients with coarctation
Aortic Coarctation

Coarctation in diastole and systole

Subclavian flap

Bypass graft

Stent (FLASH)

Stent (SSFP)

Stent (MIP)

Aortic patch with progressive aneurysm formation after 6y

Severe coarctation and collateralization

Aortic flow with severe collaterals

Index
Tetralogy of Fallot

Pre-operative findings

• VSD
• Aortic override
• RV outflow tract obstruction
• RV hypertrophy

Associated findings

• ASD
• Muscular VSD, AVSD
• PDA
• Right sided aortic arch
• Anomalous coronary arteries / pulmonary venous return
Tetralogy of Fallot

Post-operative complications

- RV outflow and/or pulmonary artery stenosis
- Pulmonary and tricuspid valve regurgitation
- RV dilation and dysfunction, LV dysfunction
- Myocardial scarring / fibrosis
- Residual ASD and VSD
- Aortic root and ascending aorta dilation
- Aortic regurgitation

Initial interventions

- BT-shunt or RVOT stent
  - If cyanosed++ neonatally
- Total repair
  - VSD patch
  - RVOT patch
  - ± conduit

Late interventions

- PV replacement (homograft, biological prosthesis)
- Re-do conduit
- PA stenting

Index
# Tetralogy of Fallot

## Protocol

1. Anatomy stack
2. VLA, HLA, LV stack, RV stack
3. LVOT, RVOT
4. PAs
5. MPA flow
6. AoV flow
7. Branch PAs flow
8. MRA PAs
9. LGE LV stack, VLA, HLA
10. 3D whole heart

## Report

1. **Size** (corrected for BSA) and **function**
   - LV: EDV, ESV, SV, EF, RWMA, (LGE)
   - RV: EDV, ESV, SV, EF, RWMA, (LGE)
2. **RVOT obstruction**: Subvalvular, valvular, supravalvular
3. **Main PA and branch PA obstruction and flows**
4. **Pulmonary regurgitation fraction** ± volume
5. **Presence** and severity of **TR**
6. **Residual shunting**: ASD, VSD, APCs; Qp : Qs
7. **Relation to coronary arteries**
8. **Aortic root and ascAo dimensions**
9. **AV regurgitation**
10. **Associated findings**
Tetralogy of Fallot

Key issues

1. **Free PR:**
   - Is common after repair of ToF
   - May be tolerated without symptoms
   - Is typically associated with a *regurgitant fraction of 35–45%*

2. **Unilateral branch PA stenosis**
   - compare LPA and RPA flow volumes

3. **Regurgitant fraction may exceed 50%, if**
   - RV is unusually large and compliant
   - Pulmonary trunk / branch PAs are large and compliant
   - Elevated pulmonary vascular resistance

4. **Late diastolic antegrade flow** in the MPA
   - Sign of restrictive RV

5. **Timing for PV replacement remains controversial. Consider:**
   - Homograft replacement may function for 15-20 years
   - Pre-operative indexed *RV EDV >160-170 mL/m²* and *RV ESV >82-85 mL/m²* fail to recover to the normal range after operation

6. **Percutaneous intervention of RVOT / branch PA obstruction**
   - Consider 3D whole heart to identify close relation to coronary arteries

Index
Tetralogy of Fallot

- RV dilation, RVOT patch, SA diastole
- RV dilatation, dyskinetic RVOT patch, SA systole
- Sagittal RVOT

- Pulmonary incompetence after RVOT patch operation
- Free PI – high blood flow in RVOT in systole – inplane
- Free PI - RVOT in diastole - inplane

Late diastolic antegrade flow in the MPA as sign of restrictive RV or severe PI
Pre-operative findings

- Underdeveloped RVOT and PV
  - Membranous PV (A)
  - Hypoplastic PAs (B)
  - Atretic PAs and MAPCAs (C)
- VSD / PDA
- Pulmonary collaterals
- RV / RA dilation and hypertrophy

Associated findings

- PFO / ASD
- Tricuspid atresia or stenosis
- dTGA / CCTGA
Pulmonary Atresia

Initial interventions

- Radiofrequency perforation of membranous PV
- BT shunt
- Total repair
  - PV valvulotomy or conduit, if suitable RV
  - Atrial septostomy and Glenn → Fontan, if RV small or coronaries depend on RV

Late interventions

- PV valvuloplasty
- PV replacement
- TV repair / replacement
- Conduit replacement
- MAPCA stenting / occlusion / unifocalization

Post-operative complications

- See BT-shunt, ToF or single ventricle physiology depending on severity and initial operation
Pulmonary Atresia

Protocol

1. Anatomy stack       6. Ao flow
2. VLA, HLA, LV stack, RV stack 7. MRA PAs
3. LVOT, RVOT              8. MRA aorta (MAPCAs)
4. PAs                       9. 3D whole heart
5. MPA ± branch PAs flow   10. LGE

Report

1. Size (corrected for BSA) and function
   • LV: EDV, ESV , SV, EF, RWMA, (LGE)
   • RV: EDV, ESV, SV, EF, RWMA, (LGE)
2. PA stenosis/ hypoplasia – extent and severity
3. MAPCAs
4. Presence and severity of VSD, PDA and / or ASD
5. Associated findings, depending on initial interventions
Key issues

1. **Pulmonary atresia vs. ToF with pulmonary atresia**
   - PAs in ToF are usually normal in size with normal peripheral arborisation
   - Systemic-to-pulmonary collaterals are less developed in ToF

2. **Complete surgical repair, if**
   - Central PAs are present
   - Sufficient PA blood supply to the lungs
   - A single PA is normal in size and reaches all lung segments

3. **Complete surgical repair is contraindicated, if**
   - Intact ventricular septum and hypoplastic right ventricle
   - Hypoplastic or absent central PAs
   - Inadequate peripheral arborization of PAs

4. **Palliative procedures**
   - BT shunt, Waterston shunt

5. **MAPCAs**
   - Are best visualized on an aortic MRA
   - Strict removal of air bubbles if MRA performed
   - Consider CT to visualize small MAPCAs
Pulmonary Atresia

- Vestigial pulmonary artery (arrow), MIP
- Major aorto-pulmonary collateral arteries - MIP

Vestigial pulmonary artery (arrow), VSD - coronal

Major aorto-pulmonary collateral arteries - MIP

Index
Dextro-Transposition of the Great Arteries - dTGA

Pre-operative findings
- VA discordance (morphological RV to aorta, LV to MPA)
- Parallel great arteries

Associated findings
- VSD / pulmonary stenosis
- LVOT obstruction (sub PS)
- PDA/ASD
- Aortic coarctation
- Coronary origin anomalies

Common interventions
- Arterial switch
- Rastelli, if accompanied with VSD and RVOT obstr. / PS
- Atrial switch – historical (Senning/Mustard)
Post-operative complications

- RVOT obstruction / MPA and branch PA stenosis
- LVOT obstruction
- Neo-aortic root dilatation
- Neo-aortic valve regurgitation
- Neo-pulmonary valve regurgitation
- Coronary artery stenosis
- Systemic RV dysfunction

Initial interventions

- Switch of aortic and pulmonary root
- Anterior positioning of distal MPA / branch PAs (LeCompte manoeuver)
- Translocation of coronary arteries from aorta to neo-aortic root

Late interventions

- AV replacement
- Ao root replacement
- RVOT enlargement
- PA stenting
# Arterial Switch

## Protocol

1. **Anatomy stack**
2. **VLA, HLA, LV stack, RV stack**
3. **LVOT, RVOT**
4. **PAs**
5. **MPA flow**
6. **AoV flow**
7. **3D whole heart**
8. **Branch PAs flow**
9. **MRA PAs**
10. **LGE**
11. **Stress myocardial perfusion**

## Report

1. **Size** (corrected for BSA) and **function**
   - LV: EDV, ESV, SV, EF, RWMA, mass
   - RV: EDV, ESV, SV, EF, RWMA
2. **RVOT / LVOT obstruction**
3. **MPA & branch PA patency (and flow)**
4. **PR and AR**
5. **Aortic dimensions**
6. **Coronary artery origins, proximal course and patency**
7. **Myocardial ischemia and / or scar**
1. **Arterial switch procedure:**
   - Is the operation of choice in dTGA
   - Is usually performed in the first two weeks of life
   - Has a favourable long-term outcome

2. **Most progressive post-operative complications:**
   - Neo-aortic regurgitation
   - Neo-pulmonary stenosis
   - Coronary obstruction

3. **If coronary obstruction is suspected, consider:**
   - 3D whole heart to assess the coronary origin and course (& patency)
   - Stress perfusion for the assessment of ischemia
   - LGE imaging for the assessment of scar
dTGA with parallel great arteries

LeCompte manoeuver with anterior PAs

Posterior PAs - side by side arteries

Supravalvular RVOT obstruction in diastole

Supravalvular RVOT obstruction in systole

RPA stenosis

Aortic root dilatation – sagittal LVOT

Aortic root dilatation – coronal LVOT

Aortic root dilation – AV stack
Post-operative complications

- Conduit or conduit valve stenosis / obstruction
- LVOT obstruction
- Residual VSD
- Residual ASD
- Branch PA stenosis

Initial interventions

- Conduit RV-MPA
- Intra-ventricular baffle
  - VSD closure
  - Redirection of left ventricular outflow to anterior aortic valve

Late interventions

- Re-operation conduit
- VSD closure device

Rastelli
Protocol

1. Anatomy stack
2. VLA, HLA, LV stack, RV stack
3. LVOT, RVOT
4. PAs
5. Conduit cross-cuts
6. MPA flow
7. AoV flow
8. Branch PAs flow
9. 3D whole heart
10. Coronal cine stack

Report

1. **Size** (corrected for BSA) and **function**
   - LV: EDV, ESV, SV, EF, RWMA, mass
   - RV: EDV, ESV, SV, EF, RWMA
2. Conduit patency and proximity to sternum
3. LVOT obstruction
4. MPA & branch PA patency (and flow)
5. Residual ASD, VSD, Qp (MPA flow) : Qs (Ao flow)
6. Course of coronary arteries and likelihood of compression if percutaneous intervention to the conduit
Key issues

1. Is usually performed between one and two years of age with a BT shunt in the meantime

2. **Allows for correction of a combination of congenital defects**
   - dTGA / double outlet right ventricle and
   - VSD and
   - RVOT obstruction
     - Pulmonary atresia
     - Pulmonary / subpulmonary stenosis

3. **Maintains systemic LV**
   - At the cost of possible LVOT obstruction and inevitable conduit interventions (surgical or percutaneous)

4. **Obstruction of RV-PA conduit**
   - The conduit runs very anteriorly, mostly directly beyond the sternum. This frequently causes an obstruction.
Intra-ventricular baffle in diastole

VSD baffle leak

RV-MPA conduit
Mustard / Senning

Post-operative complications

- Systemic and pulmonary venous baffle obstruction
- Systemic and pulmonary venous baffle leak
- Systemic RV dysfunction
- Tricuspid regurgitation
- Sub-pulmonary obstruction

Initial interventions

- Systemic venous baffle
  - directing systemic venous blood to MV
- Pulmonary venous baffle
  - directing pulmonary venous blood to TV

Late interventions

- Baffle dilation / stenting
- Closure devices
- Surgical baffle revision
- Pacemaker
## Mustard / Senning

### Protocol

1. Anatomy
2. VLA, HLA, LV stack, RV stack
3. LVOT, RVOT
4. PAs
5. Baffle cine stack in axial and SA plane
6. Baffle obstruction
7. MPA flow
8. AoV flow
9. MRA PAs
10. 3D whole heart

### Report

1. **Size** (corrected for BSA) and **function**
   - LV: EDV, ESV, SV, EF, RWMA, mass
   - RV: EDV, ESV, SV, EF, RWMA
2. Baffle obstruction
3. Baffle leak / shunt, Qp/Qs
4. Presence (and severity) of RVOT obstruction
5. Presence (and severity) of TR
Mustard / Senning

Key issues

1. **If baffle stenosis, consider**
   - 3D whole heart
   - MRA
   - Transaxial flow to assess flow reversal in azygous veins system, if SVC baffle-limb is stenosed

2. **Systemic venous baffle stenosis**
   - In 5-15% of patients, SVC > IVC
   - SVC channel patency required for transvenous pacing
   - IVC baffle stenosis less-well tolerated than SVC b. stenosis
     - Alternative blood drainage through azygos veins system
     - Elevated venous pressure on the liver

3. **Pulmonary venous baffle stenosis**
   - Physiology similar to mitral stenosis in the normal heart
   - Consider stenosis in patients with pulmonary hypertension

4. **Systemic right ventricle**
   - MRI allows longitudinal follow up and change in function
   - If dilated RV with good function and severe systemic TR valve replacement may be advantageous
   - If TR valve regurgitation due to systemic RV failure and annular dilatation then no conventional options available
Mustard / Senning

- Systemic venous baffle
- Pulmonary venous baffle
- Parallel great arteries
- Systemic RV

- Occluded SVC baffle with azygous drainage
- Reversed drainage through azygous system – same direction as flow in descAo

- Pulmonary venous baffle leak with LV volume overload
- LV (sub-pulmonary) volume overload (diastole / systole) with baffle leak
- Pulmonary venous baffle stenosis
Congenitally Corrected Transposition of the Great Arteries

Pre-operative findings
- L-TGA: AV and VA discordance
- Systemic RV
- Parallel great arteries

Associated abnormalities
- VSD
- Ebstein-like malformation of the left-sided TV
- (Sub-)pulmonary stenosis
- Aortic coarctation
- Abnormal situs
Interventions

• Depend on associated findings

Protocol

1. Anatomy
2. VLA, HLA, LV stack, RV stack
3. LVOT, RVOT
4. PAs
5. MPA flow
6. AoV flow
7. Branch PAs flow
8. 3D whole heart
9. Coronal stack
10. TR flow

Report

1. **Size** (corrected for BSA) and **function**
   • LV: EDV, ESV, SV, EF, RWMA, mass
   • RV: EDV, ESV, SV, EF, RWMA
2. **Presence and type, size and location of VSD**, jet velocity, **Qp:Qs**
3. **Presence** (and severity) of **TR**
4. **Presence** (and severity) of **(sub-)pulmonary stenosis**
5. **Other associated findings**
1. **CCTGA**
   - Usually associated with other congenital anomalies
   - A large and peri-membranous VSD is the most common associated anomaly
   - Prognosis depends on associated anomalies
   - May present late in life

2. **Coronary arteries**
   - Mirror image location

3. **Systemic RV**
   - Multiple coarse trabeculations, including the moderator band, arising from RV side of the septum
   - Best visible on RV stack (and LV stack)
   - Prone to dysfunction
   - AV valve goes with ventricle TV with RV, MV with LV
CCTGA with AV discordance

Systemic RV in diastole
Systemic RV in systole

Parallel arteries, VSD

Supravalvular RVOT stenosis in diastole
Supravalvular RVOT stenosis in systole

Congenitally Corrected Transposition of the Great Arteries
Pre-operative conditions

- Tricuspid atresia
- Double inlet ventricle
- Pulmonary atresia
- Hypoplastic left heart
- Unbalanced AVSD
- Severe Ebstein anomaly
Palliative procedure

Stage 1 - Glenn procedure

Stage 2 - Fontan completion

- Total cavo-pulmonary connection (TCPC)
  - Lateral tunnel - intracardiac
  - Extracardiac
  - Atrio-pulmonary

Additional interventions

Atrial septostomy

- to maintain systemic venous return to heart

Arterial shunt

- if inadequate pulmonary blood supply

PA banding

- if excessive pulmonary blood supply
Glenn Procedure

Post-operative complications

- Proximal insertion stenosis
- PA dilation
- Collateral formation (usually via azygous dilatation)

Initial interventions

- SVC detachment from RA
- Reconnection to RPA

Late interventions

- Usually proceeds Fontan
- Collateral vessels may require occlusion if significant desaturation
Initial interventions
- Preceded by Glenn shunt or BT shunt

Late interventions
- Occasional fenestration (arrow) closure
- Occlusion of systemic to pulmonary venous collaterals

Post-operative complications
- Stenosis to systemic venous pathways
- Ascites (due to protein losing enteropathy)
- Deterioration of ventricular function
- Thrombus always possible
- Pulmonary venous compression
- AV valve regurgitation
Fontan Procedure - Lateral Tunnel -

Initial interventions
- Preceded by Glenn shunt or BT shunt

Late interventions
- Occasional fenestration closure
- Occlusion of systemic to pulmonary venous collaterals

Post-operative complications
- Stenosis to systemic venous pathways
- Ascites (due to protein losing enteropathy)
- Deterioration of ventricular function
- Thrombus always possible
- Pulmonary venous compression
- AV valve regurgitation
Atrio-pulmonary Fontan Procedure

Post-operative complications

- Stenosis to systemic venous pathways
- Ascites (due to protein losing enteropathy)
- Deterioration of ventricular function
- Massively dilated RA
- Thrombus risk particularly high
- Pulmonary venous compression
- AV valve regurgitation
- No Glenn shunt as direct communication

Initial interventions

- Preceded by Glenn shunt or BT shunt

Late interventions

- Occasional fenestration closure
- Occlusion of systemic to pulmonary venous collaterals
**Single Ventricle Physiology**

**Protocol**

1. Anatomy stack
2. VLA, HLA, LV stack, RV stack
3. LVOT, RVOT
4. AoV
5. PAs
6. MPA ± branch PAs flow
7. AoV ± desc Ao flow
8. SC, IVC flow
9. MRA for collaterals
10. 3D whole heart
11. EGE

**Report**

1. **Size** (corrected for BSA) and **function**
   - Ventricle: EDV, ESV, SV, EF, RWMA
2. **Systemic venous pathways**
3. **PA stenosis or dilation**
4. **Presence of pulmonary vein compression**
5. **Outflow tract obstruction or dilation and VA valve function**
6. **AV valve regurgitation**
7. **Presence of thrombus**
8. **Fenestration patency**
9. **Extra cardiac findings** (pleural effusions, ascites)
Key issues

1. **Ventricular function**
   - The *single ventricle drives the circuit* so there is a *chronic low output state*
   - Deterioration in function results in worsening of clinical state in part due to *increase in LVEDP* and *the pulmonary artery pressures*

2. **AV valve function**
   - Regurgitation results in inefficiency of ventricular function, increased risk of atrial arrhythmias and increase in atrial pressures

3. **Patent systemic venous pathways**
   - If obstructed then essentially cardiac afterload increases and there is an increased risk of ascites, PLE and ventricular failure

4. **Pulmonary venous compression**
   - Particularly occurs in atrio-pulmonary Fontan as the RA dilates and compresses the right pulmonary veins.
   - Increased pulmonary venous pressure and PA pressure with the same risks as obstructed systemic venous pathways

5. **Assessment of thrombus**
   - The Fontan circuit is prone to thrombus formation due to low flow
   - If PLE – increased pulmonary vascular resistance
Single Ventricle Physiology

Lateral tunnel Fontan

Exracardiac Fontan

Classical AP Fontan with thrombus in RA

Classical AP Fontan MRA

Index
Hypoplastic Left Heart

Pre-operative findings

- Marked hypoplasia of the LV and ascending aorta
- AV and MV are atretic, hypoplastic, or stenotic
- PDA and / or ASD
- Double outlet RV in 25%

Associated findings

- Aortic coarctation
Interventions

Hybrid procedure (A)
- PDA stent, atrial septostomy, pulmonary banding

Norwood procedure (B)
Stage 1:
- MPA used to augment aorta
- RV utilised as a systemic ventricle
- PA to ascAo anastomosis to supply coronary circulation
- BT shunt or RV to PA shunt to supply PAs
- Atrial septectomy

Stage 2: Glenn procedure

Stage 3: Fontan completion

Damus-Kaye-Stancel anastomosis (augmented neo-aorta)
Post-operative complications

As per Fontan, plus:

- Aortic recoarctation
- ASD restriction / closure
- LPA stenosis
- Dysfunction of systemic RV
- TR
- Coronary insufficiency / ischemia

Protocol

1. Anatomy stack
2. VLA, HLA, LV stack, RV stack
3. LVOT, RVOT
4. AV / TV
5. PAs
6. MPA ± branch PAs flow
7. Cavo-pulm. shunt flow
8. 3D whole heart
9. MRA PAs

Report

As per Fontan, plus:

1. Neo-aortic dimensions
2. ASD flow / patency
3. Degree of TR
Key issues

Vast majority of surviving adult patients will have staged Norwood procedure and Fontan circulation. **Assessment is the same as for the Fontan circulation with additional caution:**

1. **Arch function**
   - Stenoses or dilatations where the aorta has been augmented
   - Stenosis of proximal head and neck vessels

2. **Coronary artery supply**
   - Arises from the hypoplastic ascAo which has been anastomosed to the neo-aorta
   - Careful assessment of the anastomosis

3. **AV valve function**
   - The tricuspid valve is functioning as a systemic AV valve and there is a higher likelihood that it will become regurgitant

4. **Follow-up**

Given the number of surgical interventions and the presence of a systemic ventricle which is of right morphology, it is generally accepted that these patients will, on balance, do worse than a standard Fontan in the long term.
Hypoplastic Left Heart

Hypoplastic left heart in diastole with an extracardiac Fontan

Extracardiac Fontan

Aorto-pulmonary anastomoses

Aorto-pulmonary anastomoses
Pre-operative findings

- Defects of the primum atrial septum and inlet ventricular septum
- Presence of a common atrio-ventricular valve

Associated findings

- ASD / PDA
- Coarctation of the aorta
- Anomalous pulmonary venous return
- MV anomalies, e.g. parachute MV, double orifice MV
- ToF
# Atrioventricular Septal Defect

## Interventions

- PA banding as staged approach in pulmonary overcirculation
- Surgical closure with atrial and ventricular septal patch
- Atrio-ventricular valve repair
- PDA ligation

## Post-operative complications

- Residual ASD and/or VSD
- Residual atrio-ventricular valve insufficiency or stenosis
- LVOT obstruction

## Protocol

1. **Anatomy stacks**
2. **VLA, HLA, LV stack, RV stack**
3. **LVOT, RVOT**
4. **MPA flow**
5. **Ao flow**
6. **VSD flow**
7. **ASD flow**
8. **3D whole heart**

## Report

1. **Size** (corrected for BSA) and **function**
   - LV: EDV, ESV, SV, EF, RWMA, note any LV dilation
   - RV: EDV, ESV, SV, EF, RWMA
2. **Presence and extent of ASD / VSD**
3. **Qp (MPA flow) : Qs (Ao flow)**
4. **Valve regurgitation or stenosis**
1. **Spectrum of defects**
   - Ranges from a primum ASD and cleft mitral valve to complete AVSD
   - Partial AVSD: R and L AV valves have separate orifices; usually small VSD
   - Complete AVSD: common AV valve and orifice; large VSD

2. **Most common post-operative complications requiring reoperation**
   - Severe MV regurgitation
   - LVOT obstruction

2. **Synonyms**
   - Atrioventricular canal defect
   - Endocardial cushion defect
   - Absent crux
Atrioventricular Septal Defect

AVSD - diastole

AVSD - systole

AV valve en face

AVSD – valve closure line
Pre-operative findings
• Displacement of septal/posterior tricuspid leaflet towards the apex
• Atrialization and dilation of the RV inflow
• Varying degrees of TR

Associated abnormalities
• PFO / ASD (>50%)
• VSD
• RVOT obstruction
## Interventions

- TV repair or replacement
- PFO/ASD closure
- Reduction atrioplasty
- Glenn or Fontan procedure, if severe

## Post-operative complications

- Residual tricuspid regurgitation
- RV / (LV) failure

## Protocol

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## Report

1. **Size** (corrected for BSA) and **function**
   - LV: EDV, ESV, SV, EF, RWMA
   - RV: EDV, ESV, SV, EF, RWMA
2. **Presence of PFO/ASD** or **VSD, Qp:Qs**
3. **Presence** (and severity) of **TR**
4. **Presence and severity of RVOT obstruction**
Key issues

1. The **clinical presentation** depends on the:
   - extent of tricuspid valve leaflet distortion
   - size of the right side of the heart
   - degree of TR
   - right atrial pressure
   - right-to-left shunt
   - RVOT obstruction

2. In patients with **chest pain consider myocardial ischemia** due to a compromised RCA by suture plication.

3. **Right ventricle**
   - **often dilated** despite severe apical displacement and rotation of the tricuspid valve
   - might compress the LV in diastole due to volume overload and therefore impair LV filling and **limit the cardiac output**
Ebstein Anomaly

Severe displacement of the septal tricuspid leaflet (arrow) in diastole

Severe displacement of the septal tricuspid leaflet (arrow) in systole

RV volume overload with LV compression due to diastolic septal flattening, limiting cardiac output

RV volume overload in systole
# Anomalous Coronary Arteries

## Protocol

1. **3D whole heart**

## Report

1. **Origin**
   - High / low / commissural
   - From opposite coronary sinus
   - Outside coronary sinuses
   - Separate ostium for LAD and CX
   - From pulmonary artery (ALCAPA and ARCAPA)

2. **Anomalous course**
   - Inter-arterial (anterior)
   - Retro-aortic (posterior)

3. **Anomalies of intrinsic coronary arterial anatomy**
   - Ectasia, aneurysm, hypoplasia
   - Intramural coronary artery (muscular bridge)

4. **Anomalies of coronary termination**

5. **Anomalous collateral vessels**

6. **Relation to other heart structures**, if interventions planned

---

Inter-arterial course of a LCA arising from the right sinus (A) and retro-aortic course of a LCA arising from the right sinus (B)

![Diagram](image)
Anomalous Coronary Arteries

Key issues

1. **Malignant course:**
   - Inter-arterial course between aorta and RVOT, in particular **left coronary artery from right sinus**

2. **Possible causes of ischemia:**
   - Inter-arterial dynamic compression
   - Slit-like origin
   - Myocardial bridging

3. **Consider dobutamine stress** to demonstrate a regional wall motion abnormality (if inter-arterial course), although limited prognostic value

4. **Anomalous left (or right) coronary artery from the pulmonary artery (ALCAPA/ARCAPA)**
   - Usually associated with a regional wall motion abnormality, possible infarction, MR due to affected papillary muscles or ventricular dilatation
   - Re-implantation or bypass grafting may be required if no myocardial infarction
   - Can present in adulthood
Anomalous Coronary Arteries

LCA from right sinus with inter-arterial course

RCA from left sinus with inter-arterial course

LCA from right sinus with retro-aortic course

Kinking of the proximal LCA after arterial switch procedure


<table>
<thead>
<tr>
<th>Abbreviation</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>3D</td>
<td>3 Dimensional</td>
</tr>
<tr>
<td>2CH</td>
<td>2-chamber view - VLA</td>
</tr>
<tr>
<td>3CH</td>
<td>3-chamber view</td>
</tr>
<tr>
<td>4CH</td>
<td>4-chamber view - HLA</td>
</tr>
<tr>
<td>Ao</td>
<td>Aorta</td>
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<tr>
<td>ALCAPA</td>
<td>Anomalous LCA from PA</td>
</tr>
<tr>
<td>APC</td>
<td>Atrio-pulmonary collaterals</td>
</tr>
<tr>
<td>AR</td>
<td>Aortic regurgitation</td>
</tr>
<tr>
<td>ARCAPA</td>
<td>Anomalous RCA from PA</td>
</tr>
<tr>
<td>ASD</td>
<td>Atrial septum defect</td>
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<tr>
<td>AV</td>
<td>Aortic valve</td>
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<tr>
<td>AVSD</td>
<td>Atrioventricular septal defect</td>
</tr>
<tr>
<td>BT</td>
<td>Blalock Taussig</td>
</tr>
<tr>
<td>BSA</td>
<td>Body surface area</td>
</tr>
<tr>
<td>CBTS</td>
<td>Classic BT Shunt</td>
</tr>
<tr>
<td>CCTGA</td>
<td>Congenital corrected TGA</td>
</tr>
<tr>
<td>CMR</td>
<td>Cardiac magnetic resonance</td>
</tr>
<tr>
<td>Cx</td>
<td>Circumflex artery</td>
</tr>
<tr>
<td>DCRV</td>
<td>Double chamber right ventricle</td>
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<tr>
<td>Desc</td>
<td>Descending</td>
</tr>
<tr>
<td>EDV</td>
<td>End-diastolic volume</td>
</tr>
<tr>
<td>EF</td>
<td>Ejection fraction</td>
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<tr>
<td>ESV</td>
<td>End-systolic volume</td>
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<tr>
<td>FOV</td>
<td>Field of view</td>
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<tr>
<td>HLA</td>
<td>Horizontal long axis – 4CH</td>
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<tr>
<td>HP</td>
<td>High pressure</td>
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<tr>
<td>HR</td>
<td>Heart rate</td>
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<tr>
<td>IVC</td>
<td>Inferior vena cava</td>
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<tr>
<td>LA</td>
<td>Left atrium</td>
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<tr>
<td>LAD</td>
<td>Left anterior descending artery</td>
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<tr>
<td>LCA</td>
<td>Left coronary artery</td>
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<tr>
<td>LGE</td>
<td>Late gadolinium enhancement</td>
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<tr>
<td>LPA</td>
<td>Left pulmonary artery</td>
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<tr>
<td>LV</td>
<td>Left ventricle</td>
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<tr>
<td>LVEDP</td>
<td>LV end-diastolic pressure</td>
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<tr>
<td>LVEDP</td>
<td>LV end-diastolic pressure</td>
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<td>MBTS</td>
<td>Modified BT shunt</td>
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<td>MIP</td>
<td>Maximum intensity projection</td>
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<td>MPA</td>
<td>Main pulmonary artery</td>
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<tr>
<td>MR</td>
<td>Mitral regurgitation</td>
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<tr>
<td>MRA</td>
<td>Magnetic resonance angiography</td>
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<tr>
<td>MV</td>
<td>Mitral valve</td>
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<tr>
<td>NSA</td>
<td>Number of signal averages</td>
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<tr>
<td>PA</td>
<td>Pulmonary artery</td>
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<tr>
<td>PDA</td>
<td>Persistent ductus arteriosus</td>
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<td>PFO</td>
<td>Persistent foramen ovale</td>
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<td>pHLA</td>
<td>Pseudo HLA</td>
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<tr>
<td>PI</td>
<td>Pulmonary incompetence</td>
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<tr>
<td>PLE</td>
<td>Pulmonary lung embolism</td>
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<td>PR</td>
<td>Pulmonary regurgitation</td>
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<td>PS</td>
<td>Pulmonary stenosis</td>
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<td>Pseudo SA</td>
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<tr>
<td>pVLA</td>
<td>Pseudo VLA</td>
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<td>RA</td>
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<td>RCA</td>
<td>Right coronary artery</td>
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<td>RF</td>
<td>Regurgitation fraction</td>
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<tr>
<td>RPA</td>
<td>Right pulmonary artery</td>
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<td>RV</td>
<td>Right ventricle</td>
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<td>RVOT</td>
<td>Right ventricular outflow tract</td>
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<td>RWMA</td>
<td>Regional wall motion abnormalities</td>
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<tr>
<td>SA</td>
<td>Short axis</td>
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<tr>
<td>SNR</td>
<td>Signal-to-noise ratio</td>
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<tr>
<td>SV</td>
<td>Stroke volume</td>
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<tr>
<td>SVASD</td>
<td>Sinus venous ASD</td>
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<tr>
<td>SVC</td>
<td>Superior vena cava</td>
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<tr>
<td>TCPC</td>
<td>Total cavo-pulmonary connection</td>
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<tr>
<td>TGA</td>
<td>Transposition of the great arteries</td>
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<tr>
<td>TOE</td>
<td>Transoesophageal echocardiography</td>
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<tr>
<td>ToF</td>
<td>Tetralogy of Fallot</td>
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<td>TR</td>
<td>Tricuspid regurgitation</td>
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<td>TV</td>
<td>Tricuspid valve</td>
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<tr>
<td>VENC</td>
<td>Velocity Encoding</td>
</tr>
<tr>
<td>VLA</td>
<td>Vertical long axis -2CH</td>
</tr>
</tbody>
</table>
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