Imaging in “Fontan” staged palliation

Euro Echo 2011 - CHD teaching course
Owen I Miller
Lesions?

• Staged palliation for any single ventricle lesion
  • “Fontan” for Hypoplasia of Right Heart structures
  • “Norwood” for Hypoplasia of Left Heart structures

• Left Heart
  • Mitral atresia, aortic atresia, HLHS, critical AS, Ao arch hypoplasia

• Right Heart
  • Tricuspid atresia, PA/IVS

• Unbalanced AVSD etc.
Surgical staged palliation

- **Left Heart Hypoplasia**
  1. Achieve reliable systemic blood flow
  2. D-K-S / NW 1

- **Right Heart Hypoplasia**
  1. Achieve reliable pulmonary blood flow
  2. Systemic to Pulmonary shunt if insufficient
  3. PA band if too much
Single ventricle staged palliation

- "Stage 2"
  - Superior Cavo-Pulmonary anastomosis
    - Increase pulmonary blood flow
    - Unload systemic ventricle

- "Stage 3"
  - Total Cavo-Pulmonary Connection
    - Achieve normal oxygen saturation
    - Unload systemic ventricle
Imaging for single ventricle palliation

- Diagnosis

- Surgical planning
  - Pre/post stage 1
  - Pre/post stage 2
  - Pre/post stage 3

- Ongoing surveillance

- Management of complications
Modalities

- Plain X-ray

- Echocardiography
  - Morphology
  - Functional analysis

- Angiography and hemodynamics

- MRI
  - Standard MRI
  - Hybrid MRI/Catheter ("XMR")
Inadequate left heart  “HLHS”

- Diagnostic questions:
  - Antenatal diagnosis
  - Postnatal Morphology
    - Connections
    - Pulmonary veins
    - Interatrial communication
  - HLHS vs. Borderline LV / AoV / Arch
  - Tricuspid Valve function
  - Systemic RV function
Norwood 1 operation for HLHS
“HLHS” diagnosis & pre stage 1 planning

- Plain X-ray
  - Lung volume
  - Associated contraindications, skeletal abn.

- Angiography
  - no role in initial assessment

- Cardiac MRI
  - Rarely needed for diagnosis and pre stage 1 planning
Echocardiography in pre stage 1 planning

- Morphology
- Interatrial septum
- Ventricular function
Pre-stage 1 echo assessment - TR
Borderline LV?
Borderline LV – Norwood or Hybrid?

- Modified aortic discriminant score for critical aortic stenosis (*Colon et al JACC 2006*)
  - Cut-off for Bivent. Repair (-0.46)

- Evelina Children’s Hospital Hybrids with Borderline LV
  - $N = 7$: Discriminant score: (-3.26) ~ (-1.22)
  - 3/7 survivors achieved 2 ventricle circulation
    #1 MVR @ 1 yr and Ross @ 14 months
    #2 MV repair and EFE resection @ 2 months, Ross-Konno @ 7 months
    #3 No further procedures but died @ 3 yrs after hybrid with PHT

Ballard G et Eur J Echocardiogr 2010
Hybrid procedure

- PDA Stent
- PA Bands
- BAS
Aortic discriminant score

Discriminant score

biventricular
univentricular

age (days)

Ballard G et Eur J Echocardiogr 2010
What about MRI LV volume?

Male neonate who underwent an initial coarctation repair but had a significant atrial septal defect. MRI showed a small left ventricle but very volume loaded right heart. In conjunction with his clinical condition and echocardiography, it was felt he was suitable for a biventricular repair and he underwent ASD closure and further repair of his aortic arch and is currently doing well.

Male neonate with critical aortic stenosis who underwent a balloon valvuloplasty and then a hybrid procedure. Although MRI showed a reasonably sized left ventricle, there was significant mitral regurgitation and very little forward flow through the aorta. In conjunction with his clinical condition and echocardiography, he underwent a Norwood Stage I.
Hypoplasia of Right Heart
Tricuspid Atresia
Pre stage 1 planning summary

- Predominantly echocardiographic
  - Morphology & connections
  - 2D measurements
    - Inlets
    - Outlets
    - Function
    - Regurgitation
  - MRI not routine pre stage 1
  - Angiography no longer necessary
Pre stage 2 planning

- HLHS
  S/P Norwood 1 or S/P Hybrid

  Or

- HRHS (TAT; PA/IVS)
  S/P Shunt or S/P PA Band

- Ventricular function
- Interatrial septum
- Pulmonary arteries
- SVC morphology
- Volume loading
- Outlet stenosis/regurgitation
- Shunt

- Tricuspid valve function
- Aortic arch
Function as a prognostic factor

• Initial poor RV function increases risk of late mortality
  (Altmann et al, Am J Cardiol, 2000; 86:964-8)

• Interstage mortality associated with worse function
  (Nilsson et al, Acta Paediatrica, 2006 vol. 95 (12) :1594-600)

• Absence of RV dysfunction predictive of survival to stage II
  (Walsh et al, Heart, 2009 vol. 95 (15) pp. 1238-44)
Single ventricle RV function
Subjective assessment of RV function

- Beginners
- Junior Trainees
- Echocardiographers
- Senior Trainees
- Consultants

Chart showing the percentage of concordant, 1 grade out, and 2 grades out assessments across different groups.
Tricuspid Annular Plane Systolic Excursion “TAPSE”
Speckle tracking in HLHS: Tricuspid annular descent (TMAD)

- Tricuspid/mitral annular descent
- Apical view
- Three points placed:
  - TV annulus: free wall and septum
  - Apex
- Calculates displacement of the TV plane
4 Chord Strain/Shortening

- Apical ‘4 chamber’
- Place four points:
  - TV annulus free wall
  - TV annulus septum
  - Midway down free wall
  - Midway down septal wall
- Calculates strain/shortening of each chord

Courtesy: J M Simpson / H Bellsham-Revell
Tissue Doppler & Time intervals in HLHS

RV

- Systolic and diastolic time intervals vary according to stage
- Isovolumic times appear increased
- ? Failure ? adaptation

*Figure 5*

Bellsham-Revell et al 2011 in press
MRI assessment pre stage 2

• Now in routine practice
  • Has replaced catheter angiography

• Day stay admission
  • General Anaesthesia
  • Clinical assessment and echocardiography under same GA

• Sequences
  • Cine (SSFP)
  • Black Bloods
  • Gadolinium enhanced MRA
  • 3D SSFP
cMRI – short axis stack for RV volume
Single ventricular function and arch
MRI evaluation of BT shunt
MRI – flow quantitation
MRI pre stage 2
MRI – pre HemiFontan / Glenn
Angiography pre stage 2

- Largely unnecessary
  - May be needed for shunt stenosis
  - Particularly if catheter intervention planned at same procedure

- Distal Pulmonary arteries?
  - Already seen well on MRI
  - Does not provide flow data (like MRI)
Pre stage 2 planning summary

• Predominantly cross-sectional
  • In our institution cMRI
    • Previous repair
      • Arch
      • Shunt
      • Ventricular function
      • Regurgitation
      • Pulmonary arteries

• Echocardiography still important but difficult for RV
  • Newer techniques may prove useful
# Pre stage 3 (TCPC) surgical planning: centre dependent

<table>
<thead>
<tr>
<th></th>
<th>Angiography</th>
<th>Echocardiography</th>
<th>MRI</th>
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<tbody>
<tr>
<td>Pulmonary arteries</td>
<td>+/-</td>
<td>+/-</td>
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</tr>
<tr>
<td>Aortic arch</td>
<td>✗</td>
<td>+/-</td>
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</tr>
<tr>
<td>Ventricular function</td>
<td>✗</td>
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<tr>
<td>Tricuspid Valve</td>
<td>✗</td>
<td>✓</td>
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<tr>
<td>Pulmonary veins</td>
<td>✓</td>
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Pre TCPC

- Ventricular Function
- Unobstructed Pulmonary Venous Return
- Superior cavo-pulmonary anastomosis
- Regurgitation
- Pulmonary arteries

- CVP (+/- PVRI)
FR 16Hz
11cm
2D
57%
C 47
P Low
HGen
CF
64%
2.5MHz
WF High
Med

16/06/2011 14:45:42
TIS1.5  MT 1.0
S5-1/ECH

84 bpm
MRI evaluation pre TCPC

+ CVP

Glenn

Neo-Ao

RPA

LPA
MRI pre TCPC – Volume unloading after Glenn
MRI plus Catheter for “High risk” TCPC

- If raised PVR suspected
  - then both morphology (MRI)
  - function (MRI)
  - cardiac output (MRI)
  - and invasive hemodynamics are required (Catheter)
  - May be combined with $O_2$ / iNO to complete PVRI study
XMR: combined X-ray cardiac catheterisation + Magnetic Resonance Imaging
Pre stage 3 planning summary

- Predominantly MRI + CVP
- May need formal PVRI study

For regurgitant valves:
- then 2D/3D TTE & TOE for planning valve repair
Long term follow-up after TCPC

- Patency of lateral tunnel / extra cardiac conduit
  - SEC
  - Thrombus
  - Fenestration assessment
    - Device closure

- Systemic ventricular function
  - Systemic AV valve assessment

- Veno-Venous Collaterals
Veno-venous collaterals

- Echo
  - Indirect evidence with bubble study
    - Inject onto systemic upper limb vein, image PVV return
- Angiography
  - If planning intervention
- MRI
  - Contrast enhance MRA
  - Limited by preset sequences and/or need for multiple contrast runs
- MRI-4D flow?
Angiography for V-V collaterals
MRI for V-V collateral

V-V collateral to PV
Contrast enhanced MRA vs. 4D Flow
4D flow quantification

Courtesy: I Valverde
Contrast-enhanced MRI

4D-flow MRI
Summary

• Single ventricle hearts
  • Pre stage 1
    • Comprehensive echo assessment of morphology and suitability
    • Advanced echo techniques may prove useful
  • Pre stage 2
    • Echo & MRI assessment mandatory
      • Pulmonary arteries pre Glenn / Hemi Fontan
      • Aortic arch / systemic outlet
      • Volumetry & Flows
      • Regurgitant fractions
  • Pre stage 3
    • Echo – ventricular function
    • MRI + CVP ( +/- PVRI study), morphology and resistances
Summary II

- Long term surveillance of TCPC
  - Non invasive
    - Echo:
      - Function
      - AV valve regurgitation
      - Fenestration
    - MRI:
      - Function
      - AV valve regurgitation
      - Collaterals? Role of 4D flow
      - Hybrid – XMR / PVRI studies for the “failing Fontan”
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3D TOE for device closure of fenestration
Device occlusion of TCPC fenestration