One Country-One Center: Changes in the Management of Single Ventricle Disease Stages I-III

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Department of Cardiology
Polish Mother’s Memorial Hospital
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- 250-300 interventions per year
- 400-450 surgeries per year
- Up to 40 newborns with HLHS per year
- Norwood operation is the preferred first stage
Polish Mother’s Memorial Hospital

- 250-300 interventions per year
- 400-450 surgeries per year
- Up to 40 newborns with HLHS per year
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3DRA experience:
- Introduced in 03/2010
- 479 catheterizations
- 617 3DRA runs
- 375 runs in PCPC/TCPC – 61%
3DRA in Single Ventricle Patients

• Variable and challenging anatomy
• Residual lesions
• Need for numerous studies/interventions
• Small patients, radiation exposure early in life

• Slow blood flow
• Limited wash out of contrast
• Relatively big vessels
• Small systolic-diastolic variability in vessel dimensions
- HLHS
- Norwood
- Glenn
- Fontan

Restrictive IAS

PDA stenting

Stenosis of the aortic arch

Stenosis of RV-PA shunt

Stenosis of pulmonary arteries

Veno-venous, arterio-venous collaterals, shunts

Stenosis of bidirectional Glenn shunt

Fenestration

Stenosis of extracardiac tunnel
Restrictive IAS

Stent implantation into the interatrial septum in patients with univentricular heart and a secondary restriction of interatrial communication

Tomasz Moszura, Paweł Dryńek, Sebastian Górecki, Waldemar Bobikowski, Anna Mazurek-Kula, Rafał Sarnacz, Jacek A. Moll, Alicja Siwińska, Andrzej Sysa

• Imaging for IAS stent implantation:
  • Echocardiography (TEE, TTE)
  • Angiography
HLHS

Norwood

Glenn

Fontan

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HYBRID STAGE I – PDA STENTING

Case # 2, DD

History:
15 day old male, 3.7 kg, postnatal diagnosis of critical AS at a local hospital
nCPAP -> mechanical ventilation, Prostin initiated
At the age of 3 days transferred to NICU in poor condition
Initially required Adrenaline infusion, which was later replaced with Milrinone
Echo: AA + MS
On the 8th day of life presented with fever and increase in CRP (5 g/dl; N < 1 g/dl)

Case # 2, AT

Intended intervention:
Three Dimensional Rotational Angiography
3D guided ductal stent implantation
Consideration of septostomy

Rotational Angiography:
Singlesite contrast injection
Short 6 Fr sheath in RFV
4 Fr catheter in the arterial duct
12/4 ml of 70% diluted contrast

Breath hold
1 sec delay
4.1 sec run
ROTATIONAL ANGIOGRAPHY

ORIGIN OF THE RAA

DUCTAL NARROWING

LPA

3D RECONSTRUCTION IMAGES AFTER POST-PROCESSING

3D RECONSTRUCTION OVERLAY ON FLUOROSCOPY IMAGE
STENT DEPLOYMENT – 8 X 20 MM ZILVER FLEX
12/4 ml of undiluted, 0.5 sec delay
DAP - 158 mGycm2
SUMMARY:
- Total contrast – 24 ml
- Fluoroscopy time – 6.2 min
- RA radiation dose – 355 mGycm²
- Total radiation dose – 2478 mGycm²
- RA/Total radiation dose – 0.14
- Skin in to skin out time – 35 min
METHODS AND RESULTS:

• Retrospective review of ductal stenting in 18 newborns with HLHS, including 6 patients with 3DR overlay used to guide the intervention.

• Eleven RA runs were performed, pre and post stent implantation in 5 patients and before the intervention in a single patient.

• Three-dimensional reconstructions from all RA runs had image quality sufficient to allow stent placement without additional contrast injections.

• Comparison with 2D angiography guided ductal stenting showed similar contrast usage with the 2D angiography patients receiving higher radiation dose.
CONCLUSIONS:

• Three dimensional rotational angiography provides accurate visualization of the ductal morphology and nearby structures.

• Three-dimensional reconstruction overlay with clear landing points enables precise stent implantation with no additional contrast injections and lower radiation doses than conventional angiography in our patients.
HYBRID STAGE I – VESSELNAVIGATOR GUIDED PDA STENTING

- HLHS, s/p bPAB
- 3 kg, 0.22 m²
- CT – 5 ml, 65.6 mGy cm
• **3DRA Guidance**
  - Total contrast – 24 ml
  - Fluoroscopy time – 6.2 min
  - RA radiation dose – 355 mGy cm²
  - Total radiation dose – 2478 mGy cm²
  - RA/Total radiation dose – 0.14
  - Skin in to skin out time – 35 min

• **VesselNavigator Guidance**
  - Total contrast – 5 ml
  - Fluoroscopy time – 4.3 min
  - N/A
  - Total radiation dose – 862 mGy cm²
  - N/A
  - Skin in to skin out time – 15 min
  - CT DLP – 65.6 mGy cm
- HLHS
- Norwood
- Glenn
- Fontan

- Restrictive IAS
- PDA stenting
- Stenosis of the aortic arch
- Stenosis of RV-PA shunt
- Stenosis of pulmonary arteries
- Veno-venous, arterio-venous collaterals, shunts
- Stenosis of bidirectional Glenn shunt
- Fenestration
- Stenosis of extracardiac tunnel
• HLHS, 5 months old, 5kg,
• RA with 70% stenht contrast

• Total volume 25 ml
• Injected over 5 sec
- AO 106/45/71, DAO 55/43/49, Gr – 51 mmHg
- 4 x 15 mm coronary balloon (Apex)
- 6 x 20 mm high-pressure balloon (Cordis)
- 8 x 20 mm high-pressure balloon (Cordis)
- AO 96/48/69, DAO 90/45/64, Gr – 6 mmHg
HLHS

Norwood

Glenn

Fontan

Restrictive IAS

PDA stenting

Stenosis of the aortic arch

Stenosis of RV-PA shunt

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Veno-venous, arterio-venous collaterals, shunts

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Fenestration

Stenosis of extracardiac tunnel
RV-PA Shunt and Pulmonary Arteries after NW
RV-PA Shunt and Pulmonary Arteries after NW

- 4 x 20 mm coronary balloon (Apex)
- 5 x 20 mm low-pressure balloon (Tyshak II)
HLHS
- Restrictive IAS
  - PDA stenting

Norwood
- Stenosis of the aortic arch
- Stenosis of RV-PA shunt

Glenn
- Stenosis of pulmonary arteries
  - Veno-venous, arterio-venous collaterals, shunts
  - Stenosis of bidirectional Glenn shunt

Fontan
- Fenestration
- Stenosis of extracardiac tunnel
3DRA in patients after PCPC

Rotational angiography in patients after PCPC
Sebastian Gloceny, Paweł Dryzek, Tomasz Mościcki
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Background:
Rotational angiography with three-dimensional reconstruction (3DRA) has been used to depict coronal-trabecular bone anatomy, reliable mapping and delineation of the left heart or to display the appropriate coronary sinus branch for left ventricular lead implantation. However, there has been very limited evaluation of the role of 3DRA in structural heart diseases, especially congenital heart disease, to date small numbers of studies have been conducted, in less than 10 patients.

Aim:
To evaluate the utility of rotational angiography in visualizing pulmonary vessels, qualification and planning of percutaneous interventions in patients with univentricular hearts after a second stage palliation.

Material and Methods:
We performed 3DRA (Philips Allura) in a group of 70 consecutive patients with univentricular hearts after bidirectional Glenn (BVG) and/or bidirectional atrioventricular discordance (BDAVDC) (n=70) in December 2010. All patients were diagnosed with hypoplastic left heart syndrome (HLHS) or hypoplastic left heart syndrome (HLHS) with atresia of the left ventricular outflow tract (LVOT) and/or with unbalanced AVSD (AVSD) (n=70). LVOT was defined as <25 ml/min/m² body surface area in the mitral valve area (MVA) and/or weight of 40-49 kg and weight >50 kg. We administered mean 3.3 mmol of contrast per kg of body weight. After 1.5-second interval, the mean time of fluoroscopy was 12.77 s, and time of contrast injection 5.3 s, in all patients we performed full diagnostic catheterization with balloon dilatation of stenotic vessels if required. Our 3DRA procedure was performed by an experienced cardiologist to assess the 3DRA images and choose the optimal path for percutaneous intervention. The first step of the procedure was injection of contrast into both common carotid arteries using 22G catheters. Any patient with previous coronary artery bypass graft (CABG) testing was excluded.

Two experienced pediatric interventional cardiologists who did not participate in performing of 3DRA were asked to answer questions regarding adequacy of 3DRA patients for VSD (see table).

Results:
Between 05/2010 and 12/2011, we performed 60 3DRA's in 58 patients after BVG. All patients underwent diagnostic catheterization, which in 32 (47%) was followed by 38 interventions. Median age and weight was 3.8yrs (1.5-7) and 16kg (8.5-58kg), respectively. Median contrast dose for 3DRA acquisition and for total study was 2ml/kg (7.3-3) and 4ml/kg (2.0-15.5), respectively. Median area dose for the whole study time, of fluoroscopy and total time of study was 132, cGy/cm² (25.9-1056.8), 7.7 min (0.7-80) and 52.5 min (15-180), respectively. There were no acute complications related to 3DRA. Overall quality of 3DRA images was graded by the primary operator as good in 64 (90%) and satisfactory in 9 (11%). Seven (9%) studies were graded as bad due to angiographic catheter pushed too far into the proximal pulmonary artery making visualization of superior vena cava impossible in 6 (9%), wrong localization of sources in further 2. In the remaining 3DRA's vena cava superior, Glenn connection, right and left pulmonary arteries were visualized. In all 38 interventions 3DRA images were judged by the operator to be superior to fix plane angiography in making decision concerning the interventions or in assessing the result.

Conclusions:
In patients after BVG operation 3DRA allowed for good visualization of superior vena cava, BDG shunt and course of pulmonary arteries. It was superior for fix plane angiography in planning and assessing results of percutaneous interventions.
Abstract

Patients with hypoplastic left heart syndrome require multistage surgical treatment, often supported with additional percutaneous interventions. In this population normal development of pulmonary vasculature is crucial, as it belongs to key factors reducing complication rate at all stages of palliation. Transcatheter interventions allow for significant improvement of pulmonary blood flow but they can be very challenging in the youngest patients. Three-dimensional rotational angiography (3DRA) is an emerging imaging modality that enables detailed visualization of pulmonary arteries, not achievable in standard angiography. In presented patient with univentricular heart and pulmonary artery hypoplasia 3DRA proved helpful in qualification, monitoring and final evaluation of stent implantation.

Key words: hypoplastic left heart syndrome, imaging, percutaneous intervention
• HLHS
• 2.5 years, 12 kg
• S/p NW1, BDG
• RA – 24/6 ml of plain contrast
• 2D guided Palmaz-Genesis stent (8 x 24 mm) implantation
RA and 3DR used for:
- Extensive visualization of pulmonary arteries
- Measurements (‘dynamic’ RA and ‘static’ 3DR images)
- Selection of the best angle for the intervention
- Evaluation of the final result
HLHS s/p hybrid stage I and NW + BDG

- Deferential contrast flow to the right lung
- Earlier venous return from the right lung
- Proximal stenosis to the RPA
- Part of self-expandable stent in the ligated DA
- Critical stnosis to the proximal LPA
- Diminished contrast filling of the left lung
HLHS s/p hybrid stage I and NW + BDG
RA and 3DR used for:

- Extensive visualization of pulmonary arteries
- Measurements (‘dynamic’ RA and ‘static’ 3DR images)
- Selection of the best angle for the intervention
- **Positioning of the balloon and/or stent**
- Evaluation of the final result
**Case #1, PP**

**Planar lung perfusion scintigraphy 12/2014**

Peripheral injection of $^{99m}$Tc – MAA (macroaggregated albumin)

Differential left and right lung perfusion was 8% compared with 92%

**Intended intervention:**
Three Dimensional Rotational Angiography
3D guided pulmonary artery dilation ± stent implantation

**Rotational Angiography:**
- Multi-site contrast injection
- 4 Fr Pigtail in SVC – 16/4 ml
- 4 Fr Pigtail in Neo Aorta – 16/4 ml
- 70% strength contrast
- RV rapid pacing
- Breath hold
- 1.5 sec delay
- 4.1 sec run
Rotational Angiography:
Multi-site contrast injection
4 Fr Pigtail in SVC – 16/4 ml
4 Fr Pigtail in Neo Aorta – 16/4 ml
70% strength contrast
RV rapid pacing
Breath hold
1.5 sec delay
4.1 sec run
What about the airway?
LPA vs. LMB vs. Ao
MPR – LPA vs. LMB vs. Stent in Ao
LPA vs. LMB vs. NeoAo

- Poor quality CT
- Blue rings – stent’s landing zone
- Green ring – left main bronchus
- Yellow ring – neo-aorta
- 7 x 17 mm Palamaz-Genesis stent implantation to the proximal LPA and RPA
HLHS

Norwood

Glenn

Fontan

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Fenestration

Stenosis of extracardiac tunnel
DILV + VSD + PS, 24 yo, 65 kg, After hemi-Fontan and Fontan operations
Ascites, lower extremities edema, increased Vmax in VCI
DILV + VSD + PS, 24 yo, 65 kg. After hemi-Fontan and Fontan operations. Ascites, lower extremities edema, increased Vmax in VCI.
DILV + VSD + PS, 24 yo, 65 kg, After hemi-Fontan and Fontan operations
Ascites, lower extremities edema, increased Vmax in VCI
Cordis OPTA 16 x 40 mm
CP 8z45 mm on BIB 20 x 45 mm
Visualization of the complete venous pathway in patients after Total Cavo-Pulmonary Connection
Visualization of the complete venous pathway in patients after Total Cavo-Pulmonary Connection.
Visualization of the complete venous pathway in patients after Total Cavo-Pulmonary Connection
Simultaneous Multi-Site Three Dimensional Rotational Angiography Reduces Contrast and Radiation Dose in Patients after Total Cavo-Pulmonary Connection Undergoing Catheterization

OBJECTIVE
• The aim of this study was to compare various 3DRA protocols in patients after TCPC.

METHODES
• We performed a retrospective review of all patients after TCPC who underwent 3DRA.
• Patients were assigned to three groups:
  • one single-site RA (group I – 35 patients),
  • two serial single-site RAs (group II – 11 patients)
  • simultaneous multi-site RA (group III – 16 patients).
Simultaneous Multi-Site Three Dimensional Rotational Angiography Reduces Contrast and Radiation Dose in Patients after Total Cavo-Pulmonary Connection Undergoing Catheterization

**RESULTS**

- One hundred RAs were performed in 62 TCPC patients.
- Baseline characteristics were not different amongst the groups.

<table>
<thead>
<tr>
<th></th>
<th>ALL</th>
<th>Group I</th>
<th>Group II</th>
<th>Group III</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients (%)</td>
<td>62</td>
<td>35 (56.4)</td>
<td>11 (17.8)</td>
<td>16 (25.8)</td>
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</tr>
<tr>
<td>Age (years)</td>
<td>6.7 (3.7-24)</td>
<td>7.1 (4.1-24)</td>
<td>6.5 (4.1-19.1)</td>
<td>6.4 (3.7-14.9)</td>
<td>NS</td>
</tr>
<tr>
<td>Weight (kg)</td>
<td>20 (13-80)</td>
<td>22 (13-80)</td>
<td>20 (15.5-55)</td>
<td>20 (13-58)</td>
<td>NS</td>
</tr>
<tr>
<td>BSA (m²)</td>
<td>0.83 (0.52-2)</td>
<td>0.85 (0.52-2)</td>
<td>0.84 (0.69-1.7)</td>
<td>0.8 (0.62-1.64)</td>
<td>NS</td>
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<tr>
<td>Additional 2D angiography</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>No of patients (%)</td>
<td>37 (59.7)</td>
<td>29 (82.8)</td>
<td>6 (54.5)</td>
<td>2 (12.5)</td>
<td>I vs III P&lt;0.0001</td>
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<td>II vs III P&lt;0.034</td>
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<tr>
<td>No of injections (range)</td>
<td>64 (1-5)</td>
<td>55 (1-5)</td>
<td>7 (1-2)</td>
<td>2 (1-1)</td>
<td></td>
</tr>
<tr>
<td>Contrast medium</td>
<td></td>
<td></td>
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<tr>
<td>Single RA / weight (ml/kg)</td>
<td>1.8 (0.55-2.5)</td>
<td>1.8 (0.55-2.5)</td>
<td>1.85 (1-2.1)</td>
<td>1.5 (0.6-2.2)</td>
<td>II vs III P=0.026</td>
</tr>
<tr>
<td>Total RA / weight (ml/kg)</td>
<td>2.1 (0.6-6.8)</td>
<td>2.1 (0.6-6.8)</td>
<td>4.2 (3.3-5.7)</td>
<td>1.6 (1-4.3)</td>
<td>I vs II P&lt;0.001</td>
</tr>
<tr>
<td>2D angiography / weight (ml/kg)</td>
<td>2.5 (0.4-9.1)</td>
<td>3 (0.4-9.1)</td>
<td>2.6 (0.5-7.6)</td>
<td>1.7 (0.5-4)</td>
<td>I vs III P=0.039</td>
</tr>
<tr>
<td>Total study / weight (ml/kg)</td>
<td>4.9 (1-12.6)</td>
<td>5.2 (1-11.4)</td>
<td>6.9 (4.4-12.6)</td>
<td>3.85 (1.8-5.9)</td>
<td>I vs III P=0.010 II vs III P&lt;0.001</td>
</tr>
<tr>
<td>Radiation dose</td>
<td></td>
<td></td>
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<tr>
<td>Single RA run (µGym2)</td>
<td>213.9 (99.8-1032.7)</td>
<td>233.6 (99.8-1005.0)</td>
<td>209.4 (150.6-376.3)</td>
<td>165.0 (120.0-1032.7)</td>
<td>NS</td>
</tr>
<tr>
<td>All RA runs (µGym2)</td>
<td>359.5 (135.0-2925.0)</td>
<td>342.1 (151.5-2925.0)</td>
<td>522.3 (360.0-919.3)</td>
<td>210.0 (135.0-1163.4)</td>
<td>NS</td>
</tr>
<tr>
<td>Total (µGym2)</td>
<td>814.3 (185.8-4638.3)</td>
<td>880.4 (398.8-4638.3)</td>
<td>960.9 (212.6-3449.0)</td>
<td>610.8 (185.8-1551.8)</td>
<td>I vs III P=0.0028 II vs III P=0.063</td>
</tr>
<tr>
<td>All RA share in Total dose (%)</td>
<td>44.3 (9.4-93.7)</td>
<td>31.0 (9.4-93.7)</td>
<td>68.9 (47.2-78.1)</td>
<td>45.1 (15.8-92.0)</td>
<td>NS</td>
</tr>
<tr>
<td>Fluoroscopy time (min)</td>
<td>13.3 (4.1-54.4)</td>
<td>13.4 (4.4-54.4)</td>
<td>13.3 (9.1-49.5)</td>
<td>9.5 (4.1-21.3)</td>
<td>II vs III P = 0.0568</td>
</tr>
<tr>
<td>Total study time (min)</td>
<td>70 (20-235)</td>
<td>70 (20-125)</td>
<td>70 (20-235)</td>
<td>52.5 (35-110)</td>
<td>II vs III P&lt;0.05</td>
</tr>
</tbody>
</table>
Simultaneous Multi-Site Three Dimensional Rotational Angiography Reduces Contrast and Radiation Dose in Patients after Total Cavo-Pulmonary Connection Undergoing Catheterization

RESULTS

• In group I, 29 patients (83%) received 52 additional standard 2DA,
• 6 patients (55%) had 7 additional 2DA in group II
• 2 patients (13%) had 1 additional 2DA in group III.

• Patients in group III received significantly less contrast per kg for RA, additional 2DA and total study.

• They received lower radiation dose especially when compared with those from group I and had shorter fluoroscopy and total study times.
Simultaneous Multi-Site Three Dimensional Rotational Angiography Reduces Contrast and Radiation Dose in Patients after Total Cavo-Pulmonary Connection Undergoing Catheterization

**CONCLUSIONS**

- In TCPC circulation, 3DRA with simultaneous multi-site contrast injections may contribute to *reduced radiation* and *contrast dose* when compared with single-site injection 3DRA.

- Multi-site 3DRA facilitates visualization of the entire Fontan pathway in the majority of patients and *reduces the number of adjunctive 2DA* performed.
Conclusions

- Rotational angiography clarifies anatomy of the dynamic vessels
- 3D reconstruction provides unlimited views and allows to profile any lesion
- 3D roadmapping facilitates catheterization and accelerates the interventions
- Final results of interventions are more predictable
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One Country-One Center: Changes in the Management of Single Ventricle Disease Stages I-III

THANK YOU !-)