Arterial Switch Operation
Long-term outcomes - all good?

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www.congenital-heart-disease.ch
Arterial Switch Operation
Long-term outcomes - all good?

Advisory board: Actelion, Bayer

No conflict of interest for this presentation
Transposition of the Great Arteries

all good?

- no surgery: 1955
- Baffes operation: 1955 - 1959
- atrial switch: 1960 - 1985
- arterial switch: 1985 - 2015

Birth, 1 month, 5 years, 10 years, 20 years, 40 years

historical: 1955 - 1985

worrisome: 1985 - 2015

lethal disease: 1985 - 2015

All good?
Post-operative sequelae after ASO

<table>
<thead>
<tr>
<th>Long-Term Post-Operative Sequelae</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supravalvular pulmonary stenosis*</td>
<td>~10%</td>
</tr>
<tr>
<td>Supravalvular aortic stenosis*</td>
<td>~5%</td>
</tr>
<tr>
<td>Neoaortic root dilation</td>
<td>Nearly universal</td>
</tr>
<tr>
<td>Neoaortic regurgitation</td>
<td>Most (moderate or severe in &lt;10%)</td>
</tr>
<tr>
<td>Asymptomatic coronary occlusion</td>
<td>2%-7%</td>
</tr>
<tr>
<td>Sudden cardiac death</td>
<td>&lt;1%</td>
</tr>
<tr>
<td>Arrhythmia</td>
<td>2%-10%</td>
</tr>
<tr>
<td>Aortic dissection or rupture</td>
<td>Unknown</td>
</tr>
</tbody>
</table>

*Requiring intervention. Modified with permission from Wernovsky et al. (152).
# Reported cardiac outcomes in adults after ASO

<table>
<thead>
<tr>
<th>Study</th>
<th>n</th>
<th>20y survival</th>
<th>adult deaths</th>
<th>median FU as adults</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tobler et al</td>
<td>132</td>
<td>97 %</td>
<td>0</td>
<td>3 years</td>
</tr>
<tr>
<td>JACC 2010</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Kempny et al</td>
<td>145</td>
<td>N/A</td>
<td>3 (2 cardiac)</td>
<td>9 years</td>
</tr>
<tr>
<td>Int J Cardio 2012</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Khairy et al</td>
<td>400</td>
<td>97 %</td>
<td>1 (cardiac)</td>
<td>NR</td>
</tr>
<tr>
<td>Circ 2013</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Puras et al</td>
<td>37</td>
<td>96 %</td>
<td>0</td>
<td>NR</td>
</tr>
<tr>
<td>R Esp Card 2014</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
### Reported residual lesions in adults after ASO

<table>
<thead>
<tr>
<th>Study</th>
<th>Intervention for PS</th>
<th>Aortic root dilatation</th>
<th>AR</th>
<th>AVR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tobler et al</td>
<td>3/65</td>
<td>34 mm</td>
<td>mild 52%</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td></td>
<td>25 - 48 mm</td>
<td>0 ≥ moderate</td>
<td></td>
</tr>
<tr>
<td>JACC 2010</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Kempny et al</td>
<td>11/146</td>
<td>54% dilated</td>
<td>mild 22%</td>
<td>1</td>
</tr>
<tr>
<td>Int J Cardio 2012</td>
<td></td>
<td>3pts &gt; 45 mm</td>
<td>5% ≥ moderate</td>
<td>age 17</td>
</tr>
<tr>
<td>Khairy et al</td>
<td>NR</td>
<td>31±8 mm</td>
<td>mild 47%</td>
<td>2</td>
</tr>
<tr>
<td>Circ 2013</td>
<td></td>
<td>18pts &gt; 40 mm</td>
<td>3% ≥ moderate</td>
<td>age unknown</td>
</tr>
<tr>
<td>Puras et al</td>
<td>2/37</td>
<td>16% &gt;36 mm</td>
<td>mild 22%</td>
<td>0</td>
</tr>
<tr>
<td>R Esp Card 2014</td>
<td></td>
<td></td>
<td>3% ≥ moderate</td>
<td></td>
</tr>
</tbody>
</table>
Temporal distribution of complications after ASO

Figure 2. Types of complications occurring during follow-up and temporal distribution of the complications and interventions. Ao, aorta; RVOT, right ventricular outflow tract.
## Reported cardiac status in adults after ASO

<table>
<thead>
<tr>
<th>Study</th>
<th>NYHA I</th>
<th>LVEF</th>
<th>MVO2</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Tobler et al</strong></td>
<td>100 %</td>
<td>60 %</td>
<td>73±14 %</td>
</tr>
<tr>
<td>JACC 2010</td>
<td></td>
<td>14% &lt; 55%</td>
<td></td>
</tr>
<tr>
<td><strong>Kempny et al</strong></td>
<td>92 %</td>
<td>59 %</td>
<td>NR</td>
</tr>
<tr>
<td>Int J Cardio 2012</td>
<td>3% ≥ III</td>
<td>10% &lt; 55%</td>
<td></td>
</tr>
<tr>
<td><strong>Khairy et al</strong></td>
<td>97 %</td>
<td>60 %</td>
<td>86±14 %</td>
</tr>
<tr>
<td>Circ 2013</td>
<td>0 ≥ III</td>
<td>4% &lt; 55%</td>
<td></td>
</tr>
<tr>
<td><strong>Puras et al</strong></td>
<td>93 %</td>
<td>62 %</td>
<td>NR</td>
</tr>
<tr>
<td>R Esp Card 2014</td>
<td>0 ≥ III</td>
<td>2.5% &lt; 45%</td>
<td></td>
</tr>
</tbody>
</table>
All good?
The absence of current consensus regarding the appropriate interval and modality for surveillance imaging.
Follow-up recommendations

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

at least annually

at least every 2 years

<table>
<thead>
<tr>
<th></th>
<th>FU*</th>
<th>Echo</th>
<th>BW</th>
<th>CXR</th>
<th>CPET</th>
<th>CMR/CT</th>
<th>Holter</th>
</tr>
</thead>
<tbody>
<tr>
<td>ASO</td>
<td>24</td>
<td>24</td>
<td>60</td>
<td>BL</td>
<td>BL</td>
<td>BL</td>
<td>NR</td>
</tr>
</tbody>
</table>

consensus recommendation of WATCH, not published
Surveillance imaging

Echo

LV-Function, aortic regurgitation and aortic root dilation, PA-stenosis
Surveillance imaging
CMR

LV/RV - Function/Volumes; PA’s, Coronaries, Aorta
Surveillance imaging
Ischemia?

Figure 1 Schematic demonstrating the timeline of the integrated CMR/SPECT assessment of myocardial function, perfusion, scar and coronary anatomy.

Surveillance imaging
Ischemia?

Figure 6 Example of false positive SPECT study with a fixed apical defect. (A) Steady state free precession imaging demonstrates a normal right coronary origin (solid arrow) but reveals a retro-aortic course of the left main coronary artery (dotted arrows). (B) The anomalous left main regains the normal position and bifurcates into LAD (dashed arrow) and Cx (dotted arrow). (C-D) Conventional coronary angiography confirms undistorted coronary origins (solid arrows) and substantiates the presence of an anomalous left main coronary artery (dotted arrows). (E) Bullseye plot from resting SPECT indicates a fixed apical defect suggestive of infarction. (F) Stress perfusion CMR is normal. (G, H) Late gadolinium enhancement images in 2 and 4 chamber orientations show no evidence of any apical infarction.
Surveillance imaging
Care track for ischemia

Surgical history: mode of reimplantation
Cardiovascular risk factors (BW 5y)
Clinics
ECG: Signs of Ischemia
Echo: regional wall motion, EF
CPET: MVO2, O2-Pulse
CMR baseline: branch PA, perfusion (?)
cCT baseline: coronary anatomy

WATCH, unpublished
Lack of defined management strategy when subclinical anatomic or physiologic abnormalities are identified
Adults after ASO

Challenges in lifelong management

Intima fibrosis

Aspirin? Statines? Interventions?

Pedra et al. JACC 2005
Adults after ASO

Is ASO an additional risk factor for atherosclerosis?

Asymptomatic atherosclerosis (coronary CT, Cath, IVUS)
Adults after ASO

Challenges in lifelong management

Persistent neo-aortic growth during adulthood

van der Bom et al. Heart 2014
Adults after ASO

Challenges in lifelong management

9 pts with aortic root reoperation (2.3% of ASO patients)

Figure 1: Estimated freedom from root reoperation.

Adults after ASO
Challenges in lifelong management

Aortic root aneurysm

Surgery? 5.5cm? 6.0cm? Depending on AR?

Adults after ASO
Challenges in lifelong management

Table 1: Aortic root dimensions at first reoperation after arterial switch operation

<table>
<thead>
<tr>
<th>ASO</th>
<th>Patients</th>
<th>Diagnosis</th>
<th>Age (days)</th>
<th>First aortic root procedure</th>
<th>Dimensions at First aortic root procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>BSA(^a)</td>
</tr>
<tr>
<td>1</td>
<td>TGA</td>
<td>4</td>
<td>3.0</td>
<td>AVP</td>
<td>0.93</td>
</tr>
<tr>
<td>2</td>
<td>TGA</td>
<td>2</td>
<td>7.9</td>
<td>AVR</td>
<td>0.99</td>
</tr>
<tr>
<td>3</td>
<td>TGA + VSD</td>
<td>101</td>
<td>16.0</td>
<td>AVR</td>
<td>1.87</td>
</tr>
<tr>
<td>4</td>
<td>TGA</td>
<td>3</td>
<td>8.0</td>
<td>Switch back</td>
<td>0.87</td>
</tr>
<tr>
<td>5</td>
<td>TGA</td>
<td>2</td>
<td>3.5</td>
<td>Switch back</td>
<td>0.60</td>
</tr>
<tr>
<td>6</td>
<td>TGA + VSD</td>
<td>403</td>
<td>29.0</td>
<td>SCT + MVP</td>
<td>1.79</td>
</tr>
<tr>
<td>7</td>
<td>TGA + VSD</td>
<td>6</td>
<td>20.0</td>
<td>Bentall</td>
<td>2.07</td>
</tr>
<tr>
<td>8</td>
<td>TGA</td>
<td>7</td>
<td>24.0</td>
<td>Bentall</td>
<td>1.83</td>
</tr>
<tr>
<td>9</td>
<td>TGA + VSD+PS</td>
<td>610</td>
<td>34.7</td>
<td>Bentall</td>
<td>1.70</td>
</tr>
<tr>
<td>10</td>
<td>Taussig-Bing</td>
<td>210</td>
<td>21.8</td>
<td>Bentall + VSDcl</td>
<td>1.61</td>
</tr>
<tr>
<td>11</td>
<td>Taussig-Bing</td>
<td>95</td>
<td>13.9</td>
<td>Bentall + Asc</td>
<td>1.43</td>
</tr>
<tr>
<td>12</td>
<td>TGA</td>
<td>3861</td>
<td>22.0</td>
<td>Bentall + PAPl</td>
<td>1.69</td>
</tr>
</tbody>
</table>

AR: aortic reurgitation; ARD: aortic root dilatation; ASO: arterial switch operation; Asc: ascending aorta; AVP: aortic valvuloplasty; AVR: aortic valve replacement; MR: mitral regurgitation; MVP: mitral valve plasty; PA: pulmonary artery; PAPl: pulmonary artery plasty; PAs: pulmonary artery stenosis; PS: pulmonary stenosis; PTR: pulmonary trunk reconstruction; SCT: supracoronary tube; TGA: transposition of great arteries; VSDcl: ventricular septal defect closure; ND: no data.

\(^a\)BSA: body surface area (Dubois); aortic root Z-score (Boston); aortic annulus Z-score (Boston).
Adults after ASO
Challenges in lifelong management

The rarity of symptoms attributable to potential complications (especially coronary obstruction)
Adults after ASO
Challenges in lifelong management

Sympathetic denervation of the coronary arteries with ASO
The unknown effects of acquired coronary artery disease superimposed in manipulated coronary arteries
Adults after ASO

Challenges in lifelong management

Reduced coronary blood flow reserve  Impaired myocardial blood flow under stress

Hauser et al. Circ 2001
Adults after ASO
Challenges in lifelong management

Impaired coronary reactivity

w/wo Nitroglycerin

Gagliardi et al. JACC 2005
# Behandlungsstrategie bei kardiovaskulären Risikofaktoren

<table>
<thead>
<tr>
<th>Risikofaktor</th>
<th>Interventionen/Interventionswerte (IW) / Zielwerte (ZW)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Lebensstil</strong></td>
<td></td>
</tr>
<tr>
<td>Ernährung</td>
<td>– Ausgewogene Ernährung, bedarfsgerechte Energiezufuhr</td>
</tr>
<tr>
<td></td>
<td>– Bevorzugung folgender Nahrungsmittel: Früchte, Gemüse,</td>
</tr>
<tr>
<td></td>
<td>Vollkorngetreide und -brot, fettarme Milchprodukte,</td>
</tr>
<tr>
<td></td>
<td>Fisch, mageres Fleisch</td>
</tr>
<tr>
<td></td>
<td>– Kardioprotektion: fetthaltiger Fisch und Omega-3-Fettsäuren</td>
</tr>
<tr>
<td></td>
<td>– Fett &lt; 35% der Energieaufnahme, gesättigte Fettsäuren &lt; 10% der totalen Fetteinnahme, Transfette meiden (&lt; 1%)</td>
</tr>
<tr>
<td></td>
<td>– Weniger als 5 g Kochsalz pro Tag, keine Softdrinks</td>
</tr>
<tr>
<td>Übergewicht/Adipositas</td>
<td>– Möglichst Normalgewicht anstreben und beibehalten (BMI &lt; 25)</td>
</tr>
<tr>
<td></td>
<td>– Mindestens weiteren Gewichtsanstieg vermeiden</td>
</tr>
<tr>
<td>Bewegungsmangel</td>
<td>– Hochrisiko-Patienten: Programm auf Basis eines Belastungstests; ideal 30 Minuten Ausdauertraining pro Tag</td>
</tr>
<tr>
<td></td>
<td>– Übrige Personen (Primärprävention): ideal mindestens 30 Minuten mässig intensives Ausdauertraining pro Tag und ≥ 2-mal mässig intensives Krafttraining pro Woche</td>
</tr>
<tr>
<td>Rauchen</td>
<td>– Rauchabstinenz</td>
</tr>
<tr>
<td>Alkohol</td>
<td>– Massvoller Konsum: Männer &lt; 14, Frauen &lt; 7 Standarddrinks# pro Woche</td>
</tr>
</tbody>
</table>

[www.agla.ch](http://www.agla.ch)
Adults after ASO
Anticipatory care

Low-risk lifestyle practices

- Healthy body weight
- Physically active
- No smoking
- Alcohol in moderation
- Healthy diet

Myocardial infarction
Population preventable proportions

- Healthy body weight: 79%
- Physically active: 64%
- No smoking: 44%
- Alcohol in moderation: 23%
- Healthy diet: 16%

- Blood lipids
- Blood pressure
- Oxidative stress
- Endothelial function
- Glucose tolerance
- Insulin sensitivity
- Blood coagulation
- Inflammation
- Cardiac function

Akesson et al, JACC 2014
## Adults after ASO

### Anticipatory care

#### Lipide

<table>
<thead>
<tr>
<th>Lipid</th>
<th>Risikokategorie</th>
<th>Maßnahme</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>LDL-Cholesterin</strong></td>
<td>Sehr hohes Risiko (&lt; 1.8 mmol/l (ZW)), sonst Reduktion &gt; 50%, wenn Zielwert nicht erreicht werden kann</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Hohes Risiko (&gt; 20%) (&lt; 2.5 mmol/l (ZW))</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Intermediäres Risiko (10–20%) (&lt; 3.0 mmol/l (ZW))</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Niedriges Risiko (&lt; 10%)</td>
<td>Optimierung von Lebensstil</td>
</tr>
<tr>
<td><strong>Triglyzeride</strong></td>
<td>TG &gt; 1.7 mmol/l (IW): Lebensstil Intervention, Behandlung von Grundkrankheiten und anderen Risikofaktoren</td>
<td></td>
</tr>
<tr>
<td></td>
<td>TG &gt; 2.3 mmol/l (IW) und TG &gt; 1.0 mmol/l (IW)</td>
<td>zusätzliche Medikamente</td>
</tr>
<tr>
<td></td>
<td>TG &gt; 5.0 mmol/l (IW): ev. zusätzliche Medikamente</td>
<td></td>
</tr>
<tr>
<td></td>
<td>TG &gt; 10.0 mmol/l (IW): Primärziel: Verhinderung akuter Pankreatitis, lipidologisches Konsilium</td>
<td></td>
</tr>
</tbody>
</table>

#### HDL-Cholesterin

<table>
<thead>
<tr>
<th>HDL-Cholesterin</th>
<th>Bei isoliert niedrigem HDL-Cholesterin (1.0 &lt; 1 mmol/l, TG &lt; 2.3 mmol/l): Korrektur von Lebensstilfehlern und anderer Risikofaktoren</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Bei Kombination von tiefem HDL-Cholesterin und Triglyzeride &gt; 2.3 mmol/l: Korrektur von Lebensstilfehlern und anderer Risikofaktoren, ev. zusätzliche Medikamente</td>
</tr>
</tbody>
</table>

#### Übrige Risikofaktoren

<table>
<thead>
<tr>
<th>Risikofaktor</th>
<th>Maßnahme</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Diabetes mellitus</strong></td>
<td>Blutzuckereinstellung: HbA1c &lt; 7.0%</td>
</tr>
<tr>
<td></td>
<td>LDL-C, HDL-C, TG: gemäß Risikokategorie, siehe oben Risikofaktor «Lipide»</td>
</tr>
<tr>
<td></td>
<td>Blutdruck: &lt; 140/85 mmHg (Praxis-Blutdruck, ZW)</td>
</tr>
<tr>
<td><strong>Blutdruck</strong></td>
<td>Alle Patienten: &lt; 140/90 mmHg (Praxis-Blutdruck, ZW); bei älteren Patienten mit Ausgangswerten ≥ 160 mmHg ev. Reduktion auf 140–150 mmHg</td>
</tr>
<tr>
<td><strong>Plättchenaggregation</strong></td>
<td>Primärprävention: niedrig dosierte ASS ab intermediärem Risiko oder höher</td>
</tr>
<tr>
<td></td>
<td>Sekundärprävention: Antithrombotika entsprechend klinischer Situation (siehe auch AGLA Pocketguide «Antithrombotika 2014»)</td>
</tr>
</tbody>
</table>
Adults after ASO

All good?

- almost 100% survival into adulthood after ASO
- very good long-term outcomes
  - but what about **lifelong outcomes**?
- the aortic root seems to be a real and progressive threat
- the coronary fate is still speculative
- anticipatory care (lifestyle choices, cholesterol, hypertension and exercise) is key in lifelong follow-up care
Adults after ASO

Room for improvement?

In vitro evaluation of physiological spiral anastomoses for the arterial switch operation in simple transposition of the great arteries: a first step towards a surgical alternative?

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Adults after ASO

Room for improvement?

Figure 3: The investigated different spatial relationships of the great arteries observed cranially. The aortic root is positioned 0°, 20°, 35° and 45° anterior and rightward in relation to the pulmonary root. AR: aortic root, PR: pulmonary root.
Adults after ASO

Room for improvement?

Adults after ASO

Room for improvement?

# Room for improvement?

**Surgeon volume influence on outcomes after ASO**

<table>
<thead>
<tr>
<th>Variable</th>
<th>0–2 Cases/y (n = 390)</th>
<th>3–6 Cases/y (n = 1514)</th>
<th>&gt;6 Cases/y (n = 453)</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (days)</td>
<td>7 (5–10)</td>
<td>7 (4–9)</td>
<td>6 (4–8)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Weight (kg)</td>
<td>3.4 (3.0–3.7)</td>
<td>3.4 (3.0–3.7)</td>
<td>3.3 (2.9–3.6)</td>
<td>0.002</td>
</tr>
<tr>
<td>Weight &lt;2.5 kg</td>
<td>28 (7%)</td>
<td>72 (5%)</td>
<td>43 (10%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Any preoperative risk factor</td>
<td>176 (45%)</td>
<td>777 (51%)</td>
<td>234 (52%)</td>
<td>0.09</td>
</tr>
<tr>
<td>Operative</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CPB time (min)</td>
<td>182 (156–218)</td>
<td>162 (137–192)</td>
<td>146 (117–173)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Cross-clamp time (min)</td>
<td>99 (80–123)</td>
<td>93 (77–118)</td>
<td>75 (57–89)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Length of stay (days)</td>
<td>19 (13–28)</td>
<td>18 (13–25)</td>
<td>5 (12–23)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Discharge mortality</td>
<td>26 (6.7%)</td>
<td>45 (3.0%)</td>
<td>8 (1.8%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Major complication</td>
<td>72 (18.5%)</td>
<td>223 (14.7%)</td>
<td>52 (11.5%)</td>
<td>0.02</td>
</tr>
</tbody>
</table>

Values are median and interquartile range.
CPB = cardiopulmonary bypass.
Room for improvement?

Surgeon volume influence on outcomes after ASO

Birth prevalence 0.31 (95% CI 0.28 - 0.34) per 1000 live birth
Room for improvement

Live births: 82'000
TGA: 25

http://de.statista.com/statistik/daten
Room for improvement
„The big picture“

Live births 82'000 5'235'000 150 Mio
TGA 25 1600 46'500

http://de.statista.com/statistik/daten
Thank you

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www.congenital-heart-disease.ch