Hypertension in Repaired Coarctation: When to Intervene and how to treat?

A. Eicken
Klinik für Kinderkardiologie und angeborene Herzfehler,
Deutsches Herzzentrum München,
Technische Universität München
The good news is, we repaired your baby’s coarctation...

William M. DeCampli, MD, PhD

JTCVS 2017;153:415-417

The bad news is that, despite the apparent simplicity of repair, coarctation is associated with continued risk of morbidity and decreased life expectancy, mainly stemming from the effects of chronic and often relatively refractory hypertension.
CoA is not a benign lesion!

- 90% of children with complicated coarctation died within the first year of life.
- Untreated patients surviving into adulthood usually had milder forms of coarctation.

- 25% died before age 20
- 50% died before age 32
- 75% died before age 46
- 92% died before age 60

Campbell M: 1970
Re-CoA after Surgery

What type of surgery?

A) Resection / end-end
B) Patch angioplasty
C) Interposition graft
D) Subclavian flap
E) Resection / end-end extended end-end
F) Extraanatomic bypass

(Rocchini AP, 2001)

The majority of patients were hypertensive at long-term follow-up.
• The prevalence of exercise-induced hypertension is significant, and most of these patients go on to have chronic hypertension develop.

Hager A et al. AJC 2008;101:1777-1780,

• Early repair (in infancy) may or may not reduce the risk of subsequent development of chronic hypertension.

Patients with repaired coarctation have reduced arterial response to nitroglycerin, increased carotid intima-media thickness, impaired flow-mediated dilatation, increased forearm pulse wave velocity, decreased spontaneous baroreceptor sensitivity – They have a stiff prestenotic aorta.

Luijendijk et al. IJC 2014;176:776-81;
Vriend et al. Heart 2005;91:962
De Divitiis et al. Circulation 2001;104(suppl.1):165-70
Meyer et al. EHJ 2005;26:617-22
Structural Abnormalities of Great Arterial Walls in Congenital Heart Disease
Light and Electron Microscopic Analyses

Koichiro Niwa, MD; Joseph K. Perloff, MD; Sunita M. Bhuta, MD; Hillel Laks, MD; Davis C. Drinkwater, MD; John S. Child, MD; Pamela D. Miner, NP

Background—Great arteries in congenital heart disease (CHD) may dilate, become aneurysmal, or rupture. Little is known about medial abnormalities in these arterial walls. Accordingly, we studied 18 types of CHD in patients from neonates to older adults.

Methods and Results—Intraoperative biopsies from ascending aorta, paraaortic aorta, truncus arteriosus, pulmonary trunk in 86 patients were supplemented by 16 necropsy specimens. The 142 patients were 3-88 years old (average, 32±6 years). Biopsies were examined by light (LM) and electron (EM) microscopy. Patients who had any of the following cardiac conditions were excluded: Marfan syndrome, hypoplastic left heart syndrome, endocardial cushion defects, total anomalous pulmonary venous return. Noncoronary artery disease patients and 1 transplant donor. Nine biopsies from acquired trileaflet aortic stenosis were compared with biopsies from bicuspid aortic stenosis. Negative pulmonary trunk controls were from 7 coronary disease patients. A grading system consisted of negative controls and grades 1, 2, and 3 (positive controls) by LM and EM examination of medial constituents.

Conclusions—Medial abnormalities in ascending aorta, paraaortic aorta, truncus arteriosus, and pulmonary trunk were prevalent in patients with a variety of forms of CHD encompassing a wide age range. Aortic abnormalities predispose to dilation, aneurysm, and rupture. Pulmonary trunk abnormalities may predispose to dilation, aneurysm, and rupture. Pivotal questions are whether these abnormalities are initially acquired, whether CHD plays a causal or facilitating role, and whether genetic determinants are operative. (Circulation 2001;103:393-408.)

Key Words: aorta ■ lung ■ arteries ■ heart defects, congenital

Histology: "cystic medianecrosis" with early fragmentation and fibrosis of elastic fibres and increased aortic and carotid stiffness

(Courtesy: K. Niwa, 2001)
Histology:

abnormalities in the precoarctation aortic segment (increased collagen, decreased smooth muscle and muscle content) with central aortic rigidity.

CoA in adolescents/adults

• Patients with repaired coarctation who have a so called „Gothic arch“ (one that is more angulated in its transverse portion) have a much greater prevalence of hypertension than those with a normally shaped arch

Ou P et al. JTCVS 2006:132:1105-11,
Ou P et al. EHJ 2004;25:1853-9
Aortic Arch Morphology


Gothic                   Crenel                   Normal

EuroGUCH Lausanne 2017
Aortic Arch Morphology

How successful is successful? Aortic arch shape after successful aortic coarctation repair correlates with left ventricular function

Jan L. Bruse, MSc,a Abbas Khushnood, MD,a Kristin McLeod, PhD,b,c Giovanni Biglino, PhD,a Maxime Sermesant, PhD,c Xavier Pennec, PhD,c Andrew M. Taylor, MD,a Tain-Yen Hsia, MD,a and Silvia Schievano, PhD,a for the Modeling of Congenital Hearts Alliance Collaborative Group a

3D shape models of 53 pts 22.3y post CoA repair were assessed by MRI

JTCVS 2017;153:418-427
Indications for Intervention in Coarctation of the Aorta

- All patients with a non-invasive pressure difference > 20 mmHg between upper and lower limbs, regardless of symptoms but with upper limb hypertension (> 140/90 mmHg in adults), pathologic blood pressure response during exercise, or significant LVH should have intervention.

- Independent of the pressure gradient, hypertensive patients with ≥ 50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT or invasive angiography) should be considered for intervention.

- Independent of the pressure gradient and presence of hypertension, patients with ≥ 50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT or invasive angiography) may be considered for intervention.

a = class of recommendation.  b = level of evidence.
CMR = cardiac magnetic resonance; CoA = coarctation of the aorta; CT = computed tomography; LVH = left ventricular hypertrophy.

European Heart Journal 2010; doi:10.1093/eurheartj/ehq249
Indication for treatment

• Do we need a peak systolic invasive AoA-AoD gradient > 20 mmHg to decide for treatment in an adult with arterial hypertension after CoA repair with signs of Re-CoA?

• Or should we treat more early by catheter interventional means?
Is only the gradient important?

EHP = MAP x Q
External Heart Power, Mean arterial Pressure, Q = cardiac output
IHP = myocardial wall volume x Wall stress / divided by time of Systolic contraction.

Conclusion: It was demonstrated that interventional treatment of coarctation resulted in a decrease in IHP. Pressure gradients, as the most widespread clinical parameter in CoA, did not show any correlation to changes in EHP or IHP.

Fernandes JF et al. 2017 PLOS;1-14
Abnormal Wave Reflections and Left Ventricular Hypertrophy Late After Coarctation of the Aorta Repair

Michael A. Quail, Rebekah Short, Bejal Pandya, Jennifer A. Steeden, Abbas Khushnood, Andrew M. Taylor, Patrick Segers, Vivek Muthurangu

Abstract—Patients with repaired coarctation of the aorta are thought to have increased afterload due to abnormalities in vessel structure and function. We have developed a novel cardiovascular magnetic resonance protocol that allows assessment of central hemodynamics, including central aortic systolic blood pressure, resistance, total arterial compliance, pulse wave velocity, and wave reflections. The main study aims were to (1) characterize group differences in central aortic systolic blood pressure and peripheral systolic blood pressure, (2) comprehensively evaluate afterload (including wave reflections) in the 2 groups, and (3) identify possible biomarkers among covariates associated with elevated left ventricular mass (LVM). Fifty adult patients with repaired coarctation and 25 age- and sex-matched controls were recruited. Ascending aorta area and flow waveforms were obtained using a high temporal-resolution spiral phase-contrast cardiovascular magnetic resonance flow sequence. These data were used to derive central hemodynamics and to perform wave intensity analysis noninvasively. Covariates associated with LVM were assessed using multivariable linear regression analysis. There were no significant group differences ($P\geq0.1$) in brachial systolic, mean, or diastolic BP. However central aortic systolic blood pressure was significantly higher in patients compared with controls (113 versus 107 mm Hg, $P=0.002$). Patients had reduced total arterial compliance, increased pulse wave velocity, and larger backward compression waves compared with controls. LVM index was significantly higher in patients than controls (72 versus 59 g/m², $P<0.0005$). The magnitude of the backward compression waves was independently associated with variation in LVM ($P=0.01$). Using a novel, noninvasive hemodynamic assessment, we have shown abnormal conduit vessel function after coarctation of the aorta repair, including abnormal wave reflections that are associated with elevated LVM. (Hypertension. 2017;69:501-509. DOI: 10.1161/HYPERTENSIONAHA.116.08763.) • Online Data Supplement
# Munich Experience

**Stent CoA DHM since 1999 (n = 253)**

<table>
<thead>
<tr>
<th>Gender</th>
<th>f: 87, m: 166</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median age (ys)</td>
<td>16.1 (0.01 – 63) (&gt; 14 y n = 153)</td>
</tr>
<tr>
<td>Weight (kg)</td>
<td>59.7 (2.7 – 156)</td>
</tr>
<tr>
<td>Diagnoses</td>
<td>isol. Coa 161, complex 8, other 84</td>
</tr>
<tr>
<td>CoA</td>
<td>native 109, Re-CoA 144, (Conduit 12)</td>
</tr>
<tr>
<td>Stent</td>
<td>Palmaz 13, MegaLD 28, MaxLD 32, other</td>
</tr>
<tr>
<td>Complications</td>
<td>CP 56, CP covered 72, Atrium V12/14 10</td>
</tr>
<tr>
<td>AndraXXL 6, Genesis 1, Cook formula 26</td>
<td></td>
</tr>
<tr>
<td>13/253 (5%) rupture 2 (death 1, NuDEL 1),</td>
<td></td>
</tr>
<tr>
<td>femoral comp 7, dislodgement 4,</td>
<td></td>
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<tr>
<td>aneurysm 2, compartment syndrome 1</td>
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| Diameter pre stent (mm) | 8 (0 – 10) |
| Gradient pre (mmHg) | 23 (15 – 109) |
| Gradient post (mmHg) | 4 (-14 – 24) |
Conclusion

- CoA stenting in adults is routine first line treatment in our unit
- CoA stenting in adults is associated with an increased procedural risk and you must be prepared for complications
- Long term results are unknown. The risk of hypertension is not relieved by removal of a gradient in all patients, but medical treatment is more effective
- Follow-up is mandatory (blood pressure – rest, 24h, exercise). In symptomatic patients imaging (CT or repeat cath) is indicated