Coarctation of the aorta and Coronary artery disease Cause or Result?

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History

• The first pathologic description of aortic coarctation was documented by Morgagni in 1760

• Initial operative outcomes published in 1945 by Crafoord and Gross

Aetiology

• Abnormal embryogenetic development.
• Reduced intrauterine blood flow through the aorta causing aortic underdevelopment.
• Aberrant PDA tissue in the aortic wall, which constricts the aortic lumen at the isthmus, with postnatal PDA regression.
• hey2 gene mutation resulting in VEGF upregulation (experimental-zebrafish) and NOTCH1 gene
• Syndromes: Turner, PHACE, DiGeorge, Noonan and velocardiofacial syndromes

Dijkema et al Heart 2017
Natural History

- Up to 1947 in 304 cases median age of death was 31 years with 76% of deaths attributed to aortic complications
- Heart failure, aortic rupture, bacterial endarteritis, intracranial haemorrhage, were the commonest causes of death

Jenkins and Ward Q J Med 1999
Diagnosis

- Systolic murmur at the back, suprasternal thrill
- Hypertension
- Different arterial pressure between left and right arm and right arm and legs

<table>
<thead>
<tr>
<th>Level</th>
<th>Code</th>
<th>Recommendation</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>B-NR</td>
<td>1. Initial and follow-up aortic imaging using CMR or CTA is recommended in adults with coarctation of the aorta, including those who have had surgical or catheter intervention (S4.2.6-1–S4.2.6-3).</td>
</tr>
<tr>
<td>I</td>
<td>C-EO</td>
<td>2. Resting blood pressure should be measured in upper and lower extremities in all adults with coarctation of the aorta.</td>
</tr>
<tr>
<td>IIa</td>
<td>C-LD</td>
<td>3. Ambulatory blood pressure monitoring in adults with coarctation of the aorta can be useful for diagnosis and management of hypertension (S4.2.6-4).</td>
</tr>
<tr>
<td>IIb</td>
<td>B-NR</td>
<td>4. Screening for intracranial aneurysms by magnetic resonance angiography or CTA may be reasonable in adults with coarctation of the aorta (S4.2.6-5, S4.2.6-6).</td>
</tr>
<tr>
<td>IIb</td>
<td>C-LD</td>
<td>5. Exercise testing to evaluate for exercise-induced hypertension may be reasonable in adults with coarctation of the aorta who exercise (S4.2.6-4, S4.2.6-7).</td>
</tr>
</tbody>
</table>
Imaging

- ECHO
- CT
- CMR
- Cardiac catheterization
  usually necessary for gradient measurement
3D Time Resolved Flow Imaging

van der Hulst et al J Magn Res Imag 2012
Treatment

- Surgical
- Percutaneous

<table>
<thead>
<tr>
<th>Indications</th>
<th>Class&lt;sup&gt;a&lt;/sup&gt;</th>
<th>Level&lt;sup&gt;b&lt;/sup&gt;</th>
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<tbody>
<tr>
<td>All patients with a non-invasive pressure difference $&gt;20$ mmHg between upper and lower limbs, regardless of symptoms but with upper limb hypertension ($&gt;140/90$ mmHg in adults), pathological blood pressure response during exercise, or significant LVH should have intervention</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>Independent of the pressure gradient, hypertensive patients with $\geq 50%$ aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT, or invasive angiography) should be considered for intervention</td>
<td>IIa</td>
<td>C</td>
</tr>
<tr>
<td>Independent of the pressure gradient and presence of hypertension, patients with $\geq 50%$ aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT, or invasive angiography) may be considered for intervention</td>
<td>IIb</td>
<td>C</td>
</tr>
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ESC guidelines 2010
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<tr>
<td>I</td>
<td>B-NR</td>
<td>6. Surgical repair or catheter-based stenting is recommended for adults with hypertension and significant native or recurrent coarctation of the aorta (S4.2.6-1, S4.2.6-2, S4.2.6-8–S4.2.6-12).</td>
</tr>
<tr>
<td>I</td>
<td>C-EO</td>
<td>7. GDMT is recommended for treatment of hypertension in patients with coarctation of the aorta (S4.2.6-13).</td>
</tr>
<tr>
<td>IIb</td>
<td>B-NR</td>
<td>8. Balloon angioplasty for adults with native and recurrent coarctation of the aorta may be considered if stent placement is not feasible and surgical intervention is not an option (S4.2.6-14).</td>
</tr>
</tbody>
</table>
Surgical Treatment

- Resection with End-to-End Anastomosis (abandoned due to high recoarctation rates)
- Extended resection with end to end anastomosis
- Patch aortoplasty
- Subclavian flap repair
- Coarctation Resection with Interposition Graft
- Ascending descending aorta by pass
Percutaneous Treatment

- Angioplasty since 1981
- Stenting since 1989
- Covered stents have a role in certain situations, including complex coarctation anatomy, balloon/stent related aortic perforation, or aneurysm formation post surgery or intervention
- The morbidity rate after surgical repair is greater than for primary stenting, however, the need for reintervention because of persistent hypertension appears to be greater after stent insertion.

Wilson et al. Can J Cardiol 2014
Aranson and Watkins Circulation 2015
Complications post coarctation surgical or interventional repair

<table>
<thead>
<tr>
<th>Long-term complications</th>
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</tr>
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<tbody>
<tr>
<td><strong>Local</strong></td>
<td>Recoarctation, aneurysm, dissection, rupture, fistulae, endocarditis</td>
</tr>
<tr>
<td><strong>Ascending aorta</strong></td>
<td>Aneurysm, dissection, rupture, sinus of Valsalva fistula</td>
</tr>
<tr>
<td><strong>Aortic valve</strong></td>
<td>(Bicuspid valve), stenosis, regurgitation</td>
</tr>
<tr>
<td><strong>Left ventricle</strong></td>
<td>Hypertrophy, dilation, systolic dysfunction and/or diastolic dysfunction, heart failure, sudden cardiac death</td>
</tr>
<tr>
<td><strong>Coronaries</strong></td>
<td>Premature atherosclerosis, ischaemic heart disease</td>
</tr>
<tr>
<td><strong>Cerebral</strong></td>
<td>Berry aneurysms, intracranial bleeds, atherosclerosis, stroke</td>
</tr>
<tr>
<td><strong>Systemic</strong></td>
<td>Hypertension, reduced exercise capacity</td>
</tr>
</tbody>
</table>

Dijkema et al Heart 2017
Prevalence of restenosis and dilatation in different types of repair

SSM Chen et al Int J Cardiol 2014
Coarctation of the Aorta and Coronary Artery Disease
Fact or Fiction?

- 756 coarctation patients (4.9% CAD)
- 6481 ventricular septal defect patients (3.5% CAD)

Roifman et al Circulation 2012
Patients with CoA have MI at a slightly younger age and undergo coronary intervention at a significantly younger age.

Pickard et al Am J Cardiol 2019 in Press
Subclinical Burden of Coronary Artery Calcium in Patients with Coarctation of the Aorta

- Median atherosclerotic cardiovascular disease risk score
- 8% (2-12%) for CoA patients ≥40 years
- 5% (2-9%) for patient without CoA ≥ 40 years.

Krishnamurthy et al Am J Cardiol 2018
Hypertension after coarctation repair

- Prevalence up to 32.5% post coarctation repair (range 25%-68%)
- Underlying vascular bed pathology and endothelial dysfunction is a cause for the development of hypertension despite early and anatomically successful surgical correction of the aortic coarctation (impaired FMD, PWV, distensibility)
- Factors that influence hypertension prevalence are timing of surgery, type of surgery and age at follow-up
- 64% of patients who initially had exercise-induced hypertension had gone on to develop chronic hypertension

C. Canniffe et al Int J Cardiol 2013
Recoarctation and Aneurysm formation

SSM Chen et al Int J Cardiol 2014
Contemporary late outcomes

Choudhary P et al. Heart 2015
Case

- Male, DoB 21/4/1987
- Coarctation of the aorta
- Subvalvar aortic stenosis
- Subclavian flap (Athens 1987)
- Resection subvalvar stenosis (Athens 1990)
- Balloon valvuloplasty coarctation (Athens 1993)
- Coarctation stenting 14/11/2001 (London)
- Subvalvar aortic stenosis resection (redo) 17/11/2001 (London)
- Coarctation repair with conduit 5/10/2005 (London)
Clinical Examination

- Pink, Stable
- BP RA 120/80mmHg, LA 95/70mmHg
- 2/6 systolic murmur
- Chest clear
- NT proBNP 69.5 pg/ml
- On lisinopril, bisoprolol, aspirin
ECG

- SR 55/min
- QRS duration 120ms
- IRBBB
Stress Test 2011

• He managed 8’2” (Bruce protocol)
• Under medication BP increase up to 190/100mmHg
• Max HR 140/min (66%predicted)
• No ischemic changes, no chest pain
Holter

- No significant arrhythmias recorded
MRI
ECHO
CT Angio
RCA
• We decided to start oral anticagulation
• He is well
In Conclusion

• Coarctation of the aorta is a severe disease that may be considered systemic
• Morbidity and mortality have decreased with modern surgical and percutaneous treatment
• Patients with coarctation need close life-long follow-up for hypertension and its consequences like coronary artery disease and post repair complications
• Imaging plays major role in the follow-up of these patients, and should be scheduled at least every 5 years