COARCTATION: HYPERTENSION AND EFFECT OF LONG-TERM DISEASE

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Objectives

• Define coarctation
• Discuss incidence and repair
• Discuss etiologies of hypertension
  • Pre repair
  • Post repair
• Discuss cardiac effects of long term disease
Coarctation

- Typically discrete narrowing of the proximal thoracic aorta
  - May be long-segment stenosis
  - May be associated with hypoplasia of the transverse aortic arch
  - Varies considerably in anatomy, physiology, clinical presentation, treatment options, and outcomes
- 5\textsuperscript{th} most common congenital heart defect
  - 6-8\% of live births with CHD
Pre Operative Effects

- Discrete obstruction results in afterload affect on the left ventricle
- In severe forms can result in acute heart failure, shock and acidosis in first 2 wks of life with closure of ductus arteriosus
- Less severe forms results in minimal symptoms beyond systolic upper extremity hypertension, heart murmur appreciated at the back, compensatory left ventricular hypertrophy
Long Term Unrepaired Effects

- Natural history of coarctation of the aorta. Campbell M Br Heart J 1970
- Evaluated 304 necropsies to determine natural history of coarctation patients older than 2yo

<table>
<thead>
<tr>
<th>Causes of death</th>
<th>Per cent</th>
<th>Mean age and usual decades</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congestive heart failure</td>
<td>25.5</td>
<td>39</td>
</tr>
<tr>
<td>Not directly connected</td>
<td>24</td>
<td>47</td>
</tr>
<tr>
<td>Aortic rupture</td>
<td>21</td>
<td>25</td>
</tr>
<tr>
<td>Bacterial endocarditis</td>
<td>18</td>
<td>29</td>
</tr>
<tr>
<td>Intracranial haemorrhage</td>
<td>11.5</td>
<td>29</td>
</tr>
<tr>
<td>All cases</td>
<td>100</td>
<td>34.4 years</td>
</tr>
</tbody>
</table>
TABLE I  Age at death in 304 reported necropsies

<table>
<thead>
<tr>
<th>Author</th>
<th>No. of cases</th>
<th>Percentages in each decade</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>2–9</td>
</tr>
<tr>
<td>Abbott (1928)</td>
<td>200</td>
<td>4.5</td>
</tr>
<tr>
<td>Reifenstein et al. (1947)</td>
<td>104</td>
<td>5</td>
</tr>
<tr>
<td>Total</td>
<td>304</td>
<td>4.7</td>
</tr>
</tbody>
</table>

Per cent dead at end of each decade

<table>
<thead>
<tr>
<th></th>
<th>2–9</th>
<th>10–19</th>
<th>20–29</th>
<th>30–39</th>
<th>40–49</th>
<th>50–59</th>
<th>60–69</th>
<th>70 and over</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>4.7</td>
<td>25.4</td>
<td>49</td>
<td>69.2</td>
<td>86.8</td>
<td>93.1</td>
<td>98.7</td>
<td>100</td>
</tr>
</tbody>
</table>

[Graph showing percentage of deaths by age]

[Graph showing percentage of deaths by age with two curves: one for subjects with coarctation and another for normal subjects.]
Repair

• Surgical repair conventional treatment for most children’ with coarctation of aorta
  • Extended end to end anastomosis
  • Aortic arch advancement
  • Subclavian flap aortoplasty
  • Bypass grafts between ascending and descending thoracic aorta
• Transcatheter interventions (i.e. stent or balloon angioplasty also provide additional therapeutic options
  • Use in native coarctation remains debated
Outcomes

- Early post operative outcomes excellent
  - Less than 2% early mortality
- However significant longer term morbidity remains especially with regard to the onset of premature arterial hypertension
- Coarctation likely to represent lifelong disease of arterial tree
Coarctation Long-term Assessment (COALA): Significance of arterial hypertension in a cohort of 404 patients up to 27 years after surgical repair of isolated coarctation of the aorta, even in the absence of restenosis and prosthetic material

Alfred Hager, MD, a Simone Kanz, MD, a Harald Kaemmerer, MD, VMD, FESC, a Christian Schreiber, MD, b and John Hess, MD, FESC a


- Evaluated 404 pts born before 1/1/1985 s/p surgical intervention for isolated coarctation
- 273 aged 16-73 yo underwent structured clinical evaluation with BP measurement all limbs, ambulatory BP measurement and exercise testing
- 21 (13%) had systolic brachial-ankle BP difference >20 mm Hg
Only 117 (43%) had normal BP reaction
67 (25%) already taking antihypertensive meds
63 (23%) had increased ambulatory BP
26 (10%) had BP response exceeding 2 SD of reference values
Independent risk factors for HTN were repair with prosthetic material, male sex, residual brachial-ankle BP difference, older age at follow up
Etiologies of Hypertension

• Mechanisms not fully understood
  • Recoarctation
  • Structural changes in wall of peripheral and central vessels
  • Reduced baroreceptor sensitivity
  • Alterations in renin-angiotensin system
  • Raised plasma concentrations of epinephrine and norepinephrine
  • Coexistence of essential hypertension
  • Endothelial dysfunction
• Most likely multifactorial
• Places patients at risk for premature coronary artery disease, LV hypertrophy, systolic/diastolic dysfunction
Aortic Stiffness

- Lack of aortic compliance increasing recognized as important prognostic index and potential therapeutic target in adults with hypertension
- Known to worsen as patients age and more peripherally/distal along the aorta
- Coarctation patients represent additional compliance issues
  - Suture lines, stents, and bypass grafts represent artificial areas of increased stiffness
  - Coarctation patients likely have pathology of entire arterial tree (increased rate of intracranial aneurysms ~10%)
Aortic Distensibility

- Often used in adult HTN studies
- (% Ao area $\Delta$) $\times$ Pulse pressure
- Echo studies show Ao distensibility significantly decreased compared to controls (Lombardi 2013, Brili 1998)
Aortic Pulse Wave Velocity

• Arguably remains noninvasive gold standard for arterial stiffness
• $\Delta x$ - aortic distance between two imaging levels
• $\Delta t$ - time delay between the arrival of flow
• CMR studies in adults have shown the increasing PWV in males and females as they age

Conclusion

- Coarctation simplistic description to complex anatomy, physiology, clinical presentation, treatment options, and outcomes
- Excellent short term results
- Long term issues persist with greater than 50% late term hypertension even in absence of discrete obstruction
- Coarctation of aorta should be considered lifelong disease affecting entire arterial tree
- Those with artificial material or elevated stiffness indices (i.e. PWV or aortic distensibility) likely represent the highest risk
Thank you
References


