Difficult echocardiography in an adult patient with “repaired” congenital heart disease

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Clinical history

19 year old male, 168 cm, 54 kg, BMI 19 kg/m²

Past medical history:
• Peter‘s anomaly (thinning and clouding of the cornea and attachment of the iris to the cornea, which causes blurred vision)
• At age 6: bicuspid aortic valve, mild coarctation
• At age 17: balloon dilatation of native coarctation /stent placement at the site of aortic coarctation; residual peak gradient across the stent: 2 mmHg

Regular follow-up
Clinical exam

Oxygen saturation 98% at room air

Heart rate 94 bmp and regular; normal S1, S2 with early systolic ejection click (tambour sound), 2/6 ejection murmer and 2/4 early diastolic murmer over the aorta, peripheral pulses easily palpable and not bounding, no radio-femoral delay.

Blood pressure was 146/66 mmHg right arm, 136/64 mmHg left arm, 148 mmHg systolic at the right leg (dorsalis pedis).

Medication: perindopril 5 mg OD
Question

• Based on the clinical findings, what would you expect to find at echocardiography
TTE

AORTA ABOMINALIS
TTE
TTE

IVS, d = 1.16 cm
LV, d = 4.81 cm
LVPW, d = 0.70 cm
TTE
TTE
TTE
TTE

Report:
- Bicuspid aortic valve (fused right and left-coronary cusp) with mild to moderate regurgitation, no aortic stenosis
- ascending aorta not dilated
- Peak/mean gradient at the isthmic region: 15/30 mmHg
What do you think?

• I’m confident with the clinical exam indicating no important blood pressure differences between upper and lower extremity
  ⇒ the peak gradient of 30 mmHg measured at echocardiography is not correct

• I’m confident with the echo exam indicating a > 20 mmHg blood pressure gradient across the stent at the coarctation site
  ⇒ the clinical assessment is not reliable (size BP cuff, no Doppler used for peripheral pulses, collateral vessels…)

• We need further investigations
Physics

Relationship between pressure and flow
• Conservation of energy:
  pressure = potential energy
  flow = kinetic energy

• Bernoulli equitation (fluid, steady laminar flow):
  energy density = \( P + \frac{1}{2} \rho v^2 + \rho \ g \ h = \text{constant} \)
  \( P_1 + \frac{1}{2} \rho v_1^2 + \rho \ g \ h_1 = P_2 + \frac{1}{2} \rho v_2^2 + \rho \ g \ h_2 \)
  \( P_1 - P_2 = \frac{1}{2} \rho (v_2^2 - v_1^2) \)

• Bernoulli equitation complete (fluid, pulsatile)
  \( \Delta P = \text{convective} + \text{inertial} + \text{shear stress component} \)
This means

- the simplified Bernoulli formula cannot be used to quantify pressure gradients in coarctation

- even the more complex Bernoulli equitation $P_1 - P_2 = \frac{1}{2} \rho (v_2^2 - v_1^2)$ does not take into account the viscous friction component, particularly in stented vessels

⇒ Peak echo gradients do not reflect real pressure gradients in the setting of aortic coarctation/stenting
Echo signs of significant coarctation in the adult

- Doppler flow signal isthmus - serrated pattern
  - rapid acceleration
  - early high-velocity
  - gradual deceleration throughout diastole
  (diastolic spill over)

- Doppler flow signal abdominal aorta
  - low-velocity systolic-diastolic flow

- Signs of left ventricular pressure overload:
  - LV hypertrophy
  - LV diastolic dysfunction
  - Left atrial enlargement
Echo signs of significant coarctation in the adult
Echo signs of significant coarctation in the adult
6 months later…

• presents with exertional chest pain 8/10, responding to nitroglycerin, no symptoms at rest

• physical exam: unchanged; BP right arm supine 152/78 mmHg, systolic pressure right leg 148 mmHg

• ECG unchanged;
• TTE unchanged

• Exercise ECG: left-sided stabbing chest pain beginning at mild/moderate exercise, increasing in intensity during exercise; BP peaks at 210/92 mmHg; no ECG changes
Questions – What is your next step

• increase perindopril to 10 mg OD; pain killers (paracetamol) as needed

• further investigations are needed

• no additional steps are needed – close f/u in 2 months
CT-scan
Cath
Cath
### Table 7

**Intermediate Follow-up Outcomes by Integrated Imaging**

<table>
<thead>
<tr>
<th></th>
<th>Surgery (n = 16)</th>
<th>Balloon (n = 16)</th>
<th>Stent (n = 56)</th>
<th>p Value (2-Sided)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Any complications*</td>
<td>25.0%</td>
<td>43.8%</td>
<td>12.5%</td>
<td>0.020‡</td>
</tr>
<tr>
<td>Aortic wall injury</td>
<td>12.5%</td>
<td>43.8%</td>
<td>7.1%</td>
<td>0.003‡</td>
</tr>
<tr>
<td>Dissection/intimal tear</td>
<td>0.0%</td>
<td>6.3%</td>
<td>1.8%</td>
<td>0.598</td>
</tr>
<tr>
<td>Aneurysm</td>
<td>12.5%</td>
<td>43.8%</td>
<td>5.4%</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Coarct:Dao ratio, mean</td>
<td>0.98</td>
<td>0.79</td>
<td>0.80</td>
<td>0.011‡</td>
</tr>
<tr>
<td>Coarct:Dao ratio ≥0.6</td>
<td>88%</td>
<td>93%</td>
<td>89%</td>
<td>1.000</td>
</tr>
<tr>
<td>Any reobstruction</td>
<td>18.8%</td>
<td>18.8%</td>
<td>14.3%</td>
<td>0.923</td>
</tr>
<tr>
<td>Mild†</td>
<td>6.3%</td>
<td>18.8%</td>
<td>12.5%</td>
<td></td>
</tr>
<tr>
<td>Moderate</td>
<td>6.3%</td>
<td>0%</td>
<td>1.8%</td>
<td></td>
</tr>
<tr>
<td>Severe</td>
<td>6.3%</td>
<td>0%</td>
<td>0%</td>
<td></td>
</tr>
</tbody>
</table>

*Defined as any moderate to severe reobstruction, aortic wall injury (aneurysm, dissection, intimal tear) or stent fracture. †Mild reobstruction was not considered as a complication in our analysis. ‡p < 0.05.

Coarct:Dao = narrowest coarctation dimension (mm)/the dimension of the descending aorta at the level of the diaphragm (mm).
Discussion – Echo in coarctation

• remember its physical limitations, particularly in patients with a stent

• Severity of coarctation:
  - flow pattern in descending/abdominal aorta

• Also assess
  - aortic valve and ascending aorta
  - associated lesions

• Not very sensitive for the detection of late complications after coartation repair
  - additional imaging methods are needed