The problem of aortic coarctation

Echo for selecting and monitoring procedures in congenital heart diseases

Euro Echo 2010

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Interventions in aortic coarctation

<table>
<thead>
<tr>
<th>Primary disease</th>
<th>Reinterventions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Endovascular stent</td>
<td>Endovascular stent</td>
</tr>
<tr>
<td>Balloon angioplasty</td>
<td>Balloon angioplasty</td>
</tr>
<tr>
<td>Prosthetic patch</td>
<td>Interposition graft</td>
</tr>
<tr>
<td>Homograft</td>
<td>Extraanatomic bypass</td>
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<tr>
<td>Interposition graft</td>
<td></td>
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<tr>
<td>End-to-end anastomosis</td>
<td></td>
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<tr>
<td>Subclavian flap</td>
<td></td>
</tr>
</tbody>
</table>
Interventions in aortic coarctation

www.nationwidechildren.org

James B. Seward
Endovascular approach

Surveillance of coarctation stenting

- Echo for diagnosis, prior to the intervention
- Cath: general anesthesia/moderate sedation during stent placement, aortic pressure above and below the coarctation measured before and after stent placement
- Echo, ECG (and chest Xray) d 1
- Echo after 1, 3, 6, 12 months
- Aspirin for 3 months
Terminology

• Aortic arch = portion of aorta between left common carotid and left subclavian artery

• Isthmus = portion of aorta between LSA and PDA

• Coarctation = short-segment narrowing in the region of the ligamentum arteriosum
Coarctation of the aorta

- 7% of CHD
- Males > females
- **Isolated or simple form (~82%)**
  - PDA
  - bicuspid aortic valve
- **Complex form (~18%)**
  - VSD
  - subaortric obstruction
  - mitral valve abnormalities
  - hypoplasia of arch and isthmus
  - TGA, DORV
Coarctation in Turner syndrome

- CHD in 50%
  - 30% bicuspid aortic valve
  - 12% coarctation

Coarctation and bicuspid aortic valve in Turner syndrome: most important predictors of major cardiovascular events
Williams-Beuren Syndrome

Kammache et al. J Thorac Cardiovasc Surg Nov. 2010

Supravavular AS up to 80%
Peripheral PA stenosis up to 75%
2.2% of neonates with coarctation have WB syndrome
Shone’s Complex

1. Supravalvular mitral membrane
2. Parachute mitral valve
3. Subaortic stenosis
4. Aortic coarctation
5. Bicuspid aortic valve

Usually incomplete

Coarctation of the aorta

P.O. Leary
Coarctation of the aorta

by P.O. Leary
Figure 5. Regression analysis comparing Doppler-estimated coarctation gradient (y axis), calculated using both pre- and postcoarctation velocities \[4(V_2 - V_1)\], with that measured at catheterization (CATH). All values are in millimeters of mercury.

Marx Allen. JACC 1996.
Coarctation: Doppler evaluation

- Frequent association of transverse arch hypoplasia with coarctation: increased proximal velocities

- Calculation of the systolic pressure gradient in coarctation by using the expanded Bernoulli equation $[4(V_2^2 - V_1^2)]$ if proximal velocity >1m/sec

\[ P_2 - P_1 = \text{expanded Bernoulli equation} \]

\[
\begin{align*}
V_2 &= 3.8 \text{m/sec} \\ V_2^2 &= 14.4 \\
V_1 &= 1.4 \text{m/sec} \\ V_1^2 &= 2.0
\end{align*}
\]

Without $V_1$: pressure gradient = 14.4x4= 58mmHg

With $V_1$: pressure gradient = $(14.4-2)x4=50$mmHg

P2-P1 = expanded Bernoulli equation
Problems of CW gradient

- Multiple levels or long segment of left sided obstruction: Bernoulli equation may overestimate gradient across coarctation

Giardini et al. Echocardiography 2010;27:21-31
CW gradient: caveat pulmonary stenosis

Marx Allen. JACC 1996.
Coarctation: flow abdominal aorta

P.O. Leary
Caveat PW Doppler assessment

Coarctation!

Flow celiac artery!!
Untreated coarctation in the adult

- Asymptomatic for years
- Hypertension
  UE versus LE pressure ∆
- Lower limb claudication
- Headaches
- Heart failure
- Endocarditis, endarteritis
- Aortic dissection
- Stroke or ICH

From Kpodonu et al.
Ann Thorac Surg 2010;90:1716
21-Year-Old Male with Hypertension

Mid Ascending Aorta = 54 mm

Provided by H.M. Connolly
21-Year-Old Male with Hypertension

Provided by H.M. Connolly
21-Year-Old Male with Hypertension

Descending Aorta = 1.3 m/sec
Peak 7 mmHg

Provided by H.M. Connolly
21-Year-Old Male with Hypertension

Femoral pulses very difficult to palpate
RUE BP 170, RLE BP 70 systolic

Provided by H.M. Connolly
Aortic complications in coarctation

Oliver et al. Am J Cardiol 2009; 104:1001–1006
Aortic complications in coarctation

Oliver et al. Am J Cardiol 2009; 104:1001–1006
Pseudo-aneurysms in coarctation

- Long-term anastomotic pseudoaneurysms in 3-38%
- Up to 100% rupture within 15 years
- Risk factors: prosthetic grafts, patch aortoplasty, advanced age at coarctation repair, and a hypoplastic transverse arch

„Aortic measurement is a mess“  
Daniel Murphy ACC 2010

NIH losartan trial: maximal systolic diameter, measured from inner edge to inner edge with the aortic leaflets open  
Boston Children’s data base and Z scores

ASE: views used for measurement should be those that show the largest diameter of the aortic root. No mention of systole vs. Diastole, but the figure shows leading edge to leading edge in diastole

Size aorta: www.marfan.org

- Boston data based formula (up to age 25, systole, inner to inner edge)
- Cornell data based (all ages, leading edge to leading edge, diastole)

19 year old man: 10 years after coarctation repair
Asc aortic aneurysm after coarctation repair in a 19 year old man
Complication in Turner syndrome

1996
- 16 year old, 45/XO
- 150cm, BSA 1.47m²
- Coarctation repair age 8 years (end-to-end)
- Bicuspid aortic valve, normal function
  aortic root 35mm (Z-value +4.59)
  asc aorta 30mm (Z-value +4.0), no recoarctation, „normal sized aorta“

1998
- Aortic root 36mm, BP 145/115mmHg, asc aorta 3.5
Complication in Turner syndrome

Asc aorta 3.5cm
Patient with Turner syndrome

- April 2003: age 23 → Chicken sandwich, recurrent vomiting, shivers and diarrhea during the whole night. Suddenly chest pain, nausea and weakness. GP, emergency hospitalisation. RR 150/60mmHg.
Patient with Turner syndrome

Type A aortic dissection
18 year old student with hypertension

Z score 2.35
18 year old student with hypertension
Risk factors for restenosis

- Smaller patient size
- Younger age at operation
- Era of operation
- Transverse aortic arch hypoplasia
18 year old student: after stent
Coarctation: exercise?

at rest

after 30 knee bends
Noncompaction in coarctation

20 year old man
Prior resection and end-to-end anastomosis: good result
No symptoms
Echocardiography in coarctation

- Blood pressure
- 2D imaging: aortic root, ascending aorta, associated anomalies
- M mode/LV mass
- Doppler imaging
  - aortic arch
  - descending aorta
  - abdominal aorta
  - diastolic function
Conclusion

• Echocardiography most often used imaging modality in coarctation for diagnosis and surveillance of any intervention

• Suprasternal imaging can be challenging in adults but assessment of the abdominal aorta is helpful

• Assessment of aorta and associated lesions compulsory

• Long-term complications, however, do occur and warrant lifelong follow-up
end
### Late reoperations after coarctation repair

<table>
<thead>
<tr>
<th>Procedure</th>
<th>No. of patients (% of procedures)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Recoarctation repair - patch aortoplasty, asc-to-desc aortic bypass graft</td>
<td>22 (41%)</td>
</tr>
<tr>
<td>Aortic valve surgery</td>
<td>22 (41%)</td>
</tr>
<tr>
<td>Surgery on left ventricular outflow tract</td>
<td>12 (22%)</td>
</tr>
<tr>
<td>Surgery for aneurysmal enlargement of the aorta (fals and true aneurysms)</td>
<td>6 (11%)</td>
</tr>
<tr>
<td>Mitral valve surgery</td>
<td>5 (9%)</td>
</tr>
<tr>
<td>Ventricular septal defect repair</td>
<td>4 (7%)</td>
</tr>
<tr>
<td>Coronary artery bypass grafting</td>
<td>2 (4%)</td>
</tr>
</tbody>
</table>

Polyester grafts in adult patients after coarctation repair
Aortic arch geometry after coarctation repair
Gothic or crenel?
Interrupted aortic arch

- IAA = most severe form of coarctation
- rare (1.5% of all CHD)
- Often associated with other cardiac abnormalities, including a patent ductus arteriosus, ventricular septal defect, subaortic stenosis caused by posterior malalignment of the conal septum, bicuspid aortic valve with hypoplasia of the aortic annulus, and atrial septal defect. Less commonly associated cardiac anomalies include truncus arteriosus and aortopulmonary window.
Aortic valve and aortic arch pathology after coarctation repair

Table 2 Differences between patients with and without hypertension

<table>
<thead>
<tr>
<th></th>
<th>Number of patients</th>
<th>Patients with hypertension</th>
<th>p Value</th>
</tr>
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<tbody>
<tr>
<td>Number</td>
<td>124</td>
<td>30 (24%)</td>
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</tr>
<tr>
<td>Type of surgery</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>End to end</td>
<td>91</td>
<td>19 (20%)</td>
<td>0.2220</td>
</tr>
<tr>
<td>Other</td>
<td>33</td>
<td>10 (31%)</td>
<td></td>
</tr>
<tr>
<td>Recoarctation repair</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>28</td>
<td>10 (36%)</td>
<td>0.1350</td>
</tr>
<tr>
<td>No</td>
<td>96</td>
<td>20 (21%)</td>
<td></td>
</tr>
<tr>
<td>Kinking aortic arch</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>16</td>
<td>6 (38%)</td>
<td>0.2163</td>
</tr>
<tr>
<td>No</td>
<td>108</td>
<td>24 (22%)</td>
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<tr>
<td>Cervical aortic arch</td>
<td></td>
<td></td>
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<tr>
<td>Yes</td>
<td>12</td>
<td>3 (25%)</td>
<td>1.0000</td>
</tr>
<tr>
<td>No</td>
<td>112</td>
<td>27 (24%)</td>
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<tr>
<td>Hypoplasia aortic arch</td>
<td></td>
<td></td>
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<tr>
<td>Yes</td>
<td>10</td>
<td>3 (30%)</td>
<td>0.7047</td>
</tr>
<tr>
<td>No</td>
<td>114</td>
<td>27 (24%)</td>
<td></td>
</tr>
<tr>
<td>Dilatation ascending aorta</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>35</td>
<td>11 (31%)</td>
<td>0.2553</td>
</tr>
<tr>
<td>No</td>
<td>89</td>
<td>19 (21%)</td>
<td></td>
</tr>
<tr>
<td>Aortic regurgitation</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>39</td>
<td>11 (28%)</td>
<td>0.636</td>
</tr>
<tr>
<td>No</td>
<td>85</td>
<td>20 (23%)</td>
<td></td>
</tr>
</tbody>
</table>

Roos, Hesselink et al. Heart 2003;89:1074–1077
### Aortic dissection in different populations

<table>
<thead>
<tr>
<th></th>
<th>Marfan</th>
<th>Isolated BAV</th>
<th>Coarctation</th>
<th>Turner syndrome</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Incidence population</strong></td>
<td>1:5,000</td>
<td>&gt;1:100</td>
<td></td>
<td>1:2,500 ♀</td>
</tr>
<tr>
<td><strong>Age onset, y</strong></td>
<td>20-40</td>
<td>50’s</td>
<td></td>
<td>20-40</td>
</tr>
<tr>
<td><strong>Site of dilation</strong></td>
<td>Aortic root</td>
<td>Asc aorta</td>
<td>Asc root and asc aorta</td>
<td>Asc aorta</td>
</tr>
<tr>
<td><strong>Progressive dilatation</strong></td>
<td>Yes</td>
<td>+/-</td>
<td>+/-</td>
<td>?</td>
</tr>
<tr>
<td><strong>Aortic dissection</strong></td>
<td>50 %</td>
<td>0.5 %</td>
<td>?1%?</td>
<td>1-2 %</td>
</tr>
</tbody>
</table>

Williams-Beuren Syndrome

1:10,000
Unusual facial features, short stature
IQ 40 – normal
Friendly; often anxiety disorder
Enjoy music
Hypercalcemia, premature graying of hair, diabetes, diverticulosis, cardiovascular changes

B. Pober
Coarctation: computed tomography

Collateral Circulation in Aortic Coarctation
2008 ACC/AHA ACHD guidelines

Intervention for coarctation

• Peak to peak gradient $\geq 20$ mmHg – cath data

• Peak to peak gradient $< 20$ mmHg with evidence of significant coarctation and significant collaterals
Types of coarctation
Coarctation before and after stent placement
Abdominal aortic flow

- Abdominal aortic pulse delay can be quantitated with PW Doppler by measuring the time to peak velocity in the abdominal aorta and comparing it to the same value measure from flow at the aortic annulus. This value should be indexed to heart rate, by dividing the absolute value by the square root of the RR interval. In the absence of a PDA and an early diastolic reversed flow, a corrected pulse delay value of < 2.8 is suggestive of significant coarctation (7).