Coarctation of the Aorta

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I. Embryology
   A. Affects 5% to 8% of all newborns with congenital heart disease
   B. Occurs during the 6th to 8th week of gestation
      1. Cause of Coarctation of the Aorta (CoA) is unknown. Two theories (Beekman, 2008):
         a. Ductus Tissue Theory - Postnatal constriction of aberrant ductal tissue
         b. Hemodynamic Theory - Intrauterine alterations of blood flow through the aortic arch

II. Abnormal development
   A. Deformity of the aortic isthmus (where the ductus arteriosus joins the descending aorta) – characterized by narrowing of the proximal aorta or distal to the left subclavian artery. (Moon, 2011).
      1. Localized stenosis - a shelf-like infolding of the posterior aortic wall into the aortic lumen opposite, proximal and/or distal to the ductus arteriosus (Kaemmerer, 2011)
      2. Long hypoplastic segment- a tubular hypoplasia involving the aortic arch or the aorta distal to the origin of the left subclavian artery and the ductal area (Kaemmerer, 2011)

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B. Simple CoA - coarctation in the absence of other lesions
C. Complex CoA
   1. Include intracardiac and/or extracardiac lesions (Kaemmerer, 2011):
      a. Bicuspid Aortic valve – occurs in nearly 85%, the valve may be stenotic or the annulus hypoplastic
      b. Ventricular septal defect
      c. Transposition of the great arteries
      d. Noncardiac anomaly- aneurysm of circle of Willis in 3% to 5%
D. Genetic component
   1. Turner XO syndrome- 35% of patients affected

III. Physiology
   A. Left ventricular hypertension- narrowing of the aorta causes increased resistance to left ventricular outflow therefore elevating systolic pressure
      1. Upper extremity hypertension
      2. Lower extremity BP lower than the upper body BP
      3. “Gradient” is the difference between higher upper body & decreased lower body BP
      4. Fully oxygenated arterial blood – unless other lesions are present
         a. Closure of foramen ovale & ductus arteriosus after birth causes entire cardiac output to flow through the stenotic aortic segment (Beekman, 2008)

IV. Clinical Features:
   A. Cardinal features (Kaemmerer, 2011)
      1. Upper body hypertension; weak, delayed femoral pulses; a decrease in blood pressure between upper and lower extremities; palpable collateral arteries over the medial aspect of the scapulae, the lateral chest wall, and between the ribs
      2. Thrill- suprasternal notch or neck vessels
      3. Heave- no displaced heart sound

B. Infant
   1. In severe CoA of the newborn, survival depends on patency of the ductus arteriosus
      a. When ductus arteriosus closes (approximately 8 to 10 days of life) the patient develops shock & heart failure. Metabolic disturbances, hypothermia, hypoglycemia can occur as well.
      b. Resulting in lower body & renal hypoperfusion, renal failure and/or necrotizing enterocolitis (Beekman, 2008)

C. Child or adolescent
   1. Upper extremity hypertension
      a. Widened pulse pressure as patient gets older
      b. Variability of Right and Left Arm pressures, dependent on location of CoA in relation to the left & right subclavian artery

2. Murmurs
   a. Grade 2/6 to 3/6 systolic ejection murmur at the upper left sternal border, at the base & left interscapular space (Beekman, 2008)

D. Adults
   1. Patients typically diagnosed & treated earlier in life, but may rarely present with upper extremity hypertension as an adult with a native CoA  (Daniels, 2008)

V. Medical/surgical interventions
   A. Diagnosis: most often via clinical exam, echocardiogram, and chest x-ray, MRI or CT
      1. Diagnostic cardiac catheterization (Beekman, 2008)
a. With complex anatomy and hemodynamics, of associated lesions
b. With additional clinical questions

B. Treatment & Timing: Individualized to lesion, associated conditions

1. Infant:
   a. If severe: signs occur in first hours of life
      (1) Immediate intervention required
      (2) Medical – initial stabilization, inotropic support
      (3) Prostaglandin E1 IV
         • Maintain open ductus arteriosus
         • Allows for flow from RV to enter MPA, cross the ductus, enter the aorta, and perfuse the descending aorta, renal and mesenteric arteries
   b. Surgical CoA repair
      (1) Individualized to anatomy of CoA
      (2) Plan to include treatment of any additional cardiac defects

2. Child, Adolescent
   a. Repair at 2 to 3 years of age
   b. Upon diagnosis

C. Surgery - 4 Common types of repair - regardless of technique, usually performed via a left thoracotomy incision

1. End-to-end anastomosis – 1954 by Crawford and Nylin (See illustration below)
   a. Surgical treatment of choice in most centers
   b. Excision of CoA area, circumferential anastomosis is completed with interrupted sutures anteriorly (Beekman)
2. Left subclavian flap – 1966 by Waldhausen and Nahrwold (Beekman, 2008)
   a. Ligate left subclavian artery, open the proximal subclavian artery and
      beyond the CoA
   b. Fold subclavian artery flap down over the CoA section and suture
      into place

Coarctation Repair with Left Subclavian Flap


3. Prosthetic patch aortoplasty- 1961 by Vosschulte (Beekman, 2008)
   a. Longitudinal incision is made across the CoA
   b. Area enlarged with a Dacron or Gore-tex patch

4. Bypass graft – a tube is sewn in between ascending and descending thoracic aorta

D. Outcomes
   1. Mortality rates vary on patient age and associated lesions (Kaemmerer, 2011)
      a. Simple CoA- Less than 1% mortality
      b. Age 2 to 5- best age to electively operate due to low surgical risk
      c. Death rates strongly related to complexity of lesions
d. After age 30 or 40- intraoperative mortality rate increases due to degenerative changes to the aortic wall.

2. Morbidity
   a. Post op – potential paradoxical hypertension, spinal cord ischemia & paralysis, recurrent laryngeal or phrenic nerve injury, chylothorax, bleeding, infection

E. Significant long term issues: See Section VII B

VI. Cardiac Catheterization: Interventional (Beekman, 2008)
   A. Balloon angioplasty began 1982,
      1. Initially accepted for treatment of re-coarctaton
      2. Not completely accepted for treating primary CoA
      3. Balloon Angioplasty – enlarges CoA lumen, produces linear intimal and medial tears at the CoA site

Balloon Angioplasty with Implantation of Stent


4. Risk: Aneurysm from extension to adventitia

B. Stent implantation following CoA angioplasty (See illustration above)
   1. Endovascular buttress, supports dilated aortic segment
   2. Restenosis uncommon
   3. Redilatation may be needed as child grows

C. Outcomes:
   1. Mortality- rare beyond newborn period
      a. Higher rate has been reported for angioplasty for recurrent post op CoA versus native CoA
   2. Acute complications (Beekman, 2008)
      a. Femoral artery injury and thrombosis- common in infants younger than 12 months
      b. Femoral artery hemorrhage
      c. Cerebrovascular accident
   3. Long-term problems
a. Coronary artery stenosis  
b. Dilation of neo-aortic root  
c. Aortic valve regurgitation  

VII. Long Term  
A. Excellent prognosis for normal growth, development when lesion successfully repaired in childhood (Beekman, 2008)  
B. Potential complications and risks  
1. May occur after all forms of repairs (Kaemmerer, 2011)  
2. Residual CoA – presence of gradient in aorta after repair  
3. Recurrent CoA – development of restenosis, gradient in aorta after an initially successful repair  
   a. 8% to 54% (Daniels, 2008)  
   b. Recocarctation- aortic isthmus and/or aortic arch  
   d. May cause systemic hypertension, heart failure, left ventricular wall mass, coronary artery disease  
4. Systemic arterial hypertension  
   a. At rest or during exercise (Kaemmerer, 2011)  
   b. Related to re-coarctation, structural changes in the wall of vessels, reduced baroreceptor sensitivity, changes in renin-angiotensin system, elevated plasma levels of epinephrine and norepinephrine, or coexistence of essential hypertension  
   c. Higher risk of residual hypertension & early atherosclerotic disease if repairs occur late in childhood and adolescence (Maron, 1973)  
   d. Systolic hypertension may occur during dynamic exercise (Beekman, 2008)  
5. Coronary artery disease  
6. Progression valve disease, bicuspid aortic valve or mitral valve (Daniels, 2008a)  
   a. Bicuspid aortic valve can progress to stenosis (59-81%) or regurgitation (13-22%) (Sabet, 1999)  
7. Aortic aneurysm at the site of CoA, ascending or descending aorta  
   a. Highest after prosthetic patch aortoplasty  
   b. Risk rupture (Kaemmerer, 2011) – dangerous, life threatening  
8. Other vascular problems: Aortic dissection, intracranial hemorrhage (may be related to berry aneurysms in circle of Willis) (Beekman, 2008)  
9. Long term concerns may be greatly affected by associated cardiac lesions  
10. Left shoulder elevation- seen in adults due to left lateral thoracotomy  
11. Left arm – decreased pulse/ BP if surgery used left subclavian artery patch  
12. Sudden death (Daniels, 2008)  
13. Bacterial endocarditis  
   a. Antibiotic prophylaxis prior to dental procedures no longer required by American Heart Association, 2007  
   b. Should seek additional information regarding status of other lesions  

VIII. Adults with CoA repair: Routine cardiology care  
A. Annual visits: Complex congenital heart disease classification (Kaemmerer, 2011)  
   1. Clinical evaluation  
      a. Documentation of type of CoA repair is important  
      b. Monitor blood pressures  
      c. Assessment NOTE: Using four extremities BP needed  
         (1) Monitor for upper body hypertension, weak femoral pulses, gradient between upper and lower body BP
(2) If left subclavian artery used as part of repair, BP’s will be LOWER in the left arm (avoid use of arterial line here)
(3) If aberrant subclavian artery present – must consider use of left arm to obtain a BP which is proximal to CoA repair

d. Electrocardiogram
e. Transthoracic echo
f. Cardiac magnetic resonance (MRI) or CT
   (1) Surveillance aorta for potential aneurysms, status of aortic repair, valves
g. Exercise test: Surveillance for exercise induced hypertension
h. Monitor new or different type headache
   (1) May be sign of possible cerebral aneurysm
i. Monitor cholesterol; avoid obesity & smoking
   (1) Minimize additional risk for coronary artery disease
j. Education
   (1) Assess knowledge, review condition, life long care needs
      (Resources: American Heart Association (americanheart.org),
      Adult Congenital Heart Association (www.achaheart.org))
   (2) Pregnancy information website:
      http://www.heartdiseaseandpregnancy.com/

B. Care during pregnancy (Refer to problem section on pregnancy in adults with CHD
   1. Recommendations
      a. Consultation: Adult congenital heart cardiologist before pregnancy
      b. Multidisciplinary observation by experienced cardiologist during pregnancy, labor, delivery and for period of time post-partum
      c. Highest risks
         (1) Unrepaired CoA, arterial hypertension, residual CoA, aneurysm
             (Kaemmerer, 2011)

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