Coarctation of the Aorta
What the Nurse Caring for a Patient with Congenital Heart Disease Needs to Know

Jo Ann Nieves, MSN, ARNP, CPN, PNP-BC, FAHA
Nurse Practitioner, Adult Congenital Heart Program
Nicklaus Children’s Hospital
Miami, Florida

Amanda Green, MSN, ARNP, FNP-C
Nurse Practitioner, Cardiac Catheterization
Nicklaus Children’s Hospital
Miami, Florida

Embryology
• Affects 5% to 8% of all newborns with congenital heart disease (Krieger, 2015)
• Occurs during the 6th to 8th week of gestation
  o Cause of Coarctation of the Aorta (CoA) is unknown; there are two theories as to the causation of coarctation (Beekman, 2008):
    ▪ Ductus Tissue Theory- Postnatal constriction of aberrant ductal tissue
    ▪ Hemodynamic Theory- Intrauterine alterations of blood flow through the aortic arch

Abnormal development
• 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).
  o 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)
  o Long hypoplastic segment- a tubular hypoplasia involving the aortic arch or the aorta distal to the origin of the left subclavian artery and the ductus area (Kaemmerer, 2011)

- Simple CoA: coarctation in the absence of other lesions
- Complex CoA (Krieger, 2015)
  - Includes intracardiac and/or extracardiac lesions
    - Bicuspid Aortic valve – occurs in 50-60%
    - Ventricular septal defect, atrial septal defect
  - CoA & Complex CHD (Transposition of the great arteries, atrioventricular canal defect, hypoplastic left heart syndrome)
    - CoA can present with other forms of left heart obstruction (mitral stenosis, subaortic stenosis, aortic stenosis)
    - Noncardiac anomaly- intracranial aneurysm (10%)
    - Of those patients with a bicuspid aortic valve, 5% of those patients will also have CoA
- Genetic component
  - In Turner XO syndrome - 2 35% of patients have CoA (Krieger, 2015)

**Physiology**
- Left ventricular hypertension
  - Narrowing of the aorta causes increased resistance to left ventricular outflow resulting in elevated systolic pressure
  - Upper extremity hypertension (Krieger, 2015)
  - Lower extremity BP lower than the upper body BP
  - “Gradient” is the difference between higher upper body & decreased lower body BP
- Closure of ductus arteriosus
  - Results in fully oxygenated arterial blood – unless other lesions are present
  - Closure of foramen ovalae and ductus arteriosus after birth causes entire cardiac output to flow through the stenotic aortic segment (Beekman, 2008)

**Clinical Features**
- Cardinal features (Krieger, 2015; Kaemmerer, 2011)
  - Upper body arterial hypertension
  - Weak, absent, and/or delayed femoral pulses
  - Decrease in blood pressure in lower extremities
  - Palpable collateral arteries over the medial aspect of the scapulae, the lateral chest wall, and between the ribs
    - Thrill- suprasternal notch or neck vessels
    - Heave- no displaced heart sound
- Infant
  - Severe CoA of the newborn
    - Survival depends on patency of the ductus arteriosus
    - When ductus arteriosus closes (approximately 8 to 10 days of life
• Newborn develops:
  o Shock & heart failure
  o Metabolic disturbances
  o Hypothermia
  o Hypoglycemia

  ▪ Results in: (Beekman, 2008)
    • Lower body
    • Renal hypoperfusion with renal failure
    • Necrotizing enterocolitis (NEC)

• Child or adolescent
  o Upper extremity hypertension
    ▪ Widened pulse pressure as patient gets older
    ▪ Variability of Right and Left Arm pressures, dependent on location of
      CoA in relation to the left & right subclavian artery
  o Murmurs
    ▪ Grade 2/6 to 3/6 systolic ejection murmur at the upper left sternal border,
      at the base & left interscapular space posteriorly (Beekman, 2008)

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

Medical/surgical interventions
• Diagnosis:
  o Most often via clinical exam, echocardiogram, and chest x-ray, MRI or CT
  o Diagnostic cardiac catheterization only if anatomy and hemodynamics, associated
    lesions are more complex, and additional clinical questions are present (Beekman,
    2008)

• Treatment & Timing:
  o Individualized to lesion, associated conditions
  o Infant: If severe CoA, signs occur in first hours of life
    ▪ Immediate intervention required
      ▪ Medical – initial stabilization, inotropic support
      ▪ Prostaglandin E1 IV - maintain open ductus arteriosus
        o Allows for flow from RV to enter MPA, cross the ductus,
          enters the aorta & perfuses the descending aorta, renal &
          mesentery arteries
    ▪ Surgical CoA repair
    ▪ May require individualized plan to treat any additional cardiac defects
  o Child, adolescent
    ▪ Repair at 2 to 3 years of age, or upon diagnosis
  o Adult
    ▪ In adults, endovascular stenting by cardiac cath has largely supplanted
      traditional surgery (Bhatt, 2015)
Surgery - 4 Common types of repair - regardless of technique, usually performed via a left thoracotomy incision
  o End to end anastomosis – 1945 (Vonder Muhull, 2016)
    ▪ Surgical treatment of choice in most centers
    ▪ Excision of CoA area, circumferential anastomosis is completed with interrupted sutures anteriorly (Beekman, 2008)

  o Left subclavian flap – 1966 by Waldhausen and Nahrwold (Beekman, 2008)
    ▪ Ligate left subclavian artery, open the proximal subclavian artery and beyond the CoA
    ▪ Subclavian artery flap is folded down over the CoA section and sutured into place
Coarctation Repair with Left Subclavian Flap

- Prosthetic patch aortoplasty - 1961 by Vosschulte (Beekman, 2008)
  - Longitudinal incision is made across the CoA
  - Area enlarged with a Dacron or Gore-Tex® patch
- Bypass graft
  - A tube is sewn in between the ascending & descending thoracic aorta
- Outcomes
  - Mortality rates vary on patient age and associated lesions (Kaemmerer, 2011)
    - Simple CoA - Low mortality: Neonate 2.1%; Infant 0.64%; Child 0% (STS.org, 2016)
    - Age 2 to 5 - best age to electively operate due to low surgical risk
    - Death rates strongly related to complexity of any additional lesions
    - Rarely diagnosed in adults > 40 year old (Bhatt, et al., 2015).
      Untreated CoA has 75% mortality by age 46 years (Bhatt, et al., 2015)
    - After age 30 or 40 - intraoperative mortality rate increases due to degenerative changes to the aortic wall.
  - Morbidity
    - Post-operative risks:
      - Potential paradoxical hypertension
- Spinal cord ischemia & paralysis
- Recurrent laryngeal or phrenic nerve injury
- Chylothorax
- Bleeding
- Infection

- Significant long term issues: See Section on Long Term Care below

- Cardiac Catheterization: Interventional, Balloon angioplasty, potential stent
  - Balloon angioplasty
    - Began 1982
    - Widely accepted for treating recoarctaton
    - Enlarges CoA lumen
      - Produces linear intimal and medial tears at the CoA site
      - Artery tear may extend to adventitia – risk aneurysm
  - Stent implantation following CoA angioplasty (See illustration below)

![](Balloon%20Angioplasty%20with%20Implantation%20of%20Stent.png)

Balloon Angioplasty with Implantation of Stent
© Scientific Software Solutions, 2016. All rights reserved.

- Endovascular buttress, supports the arterial wall and opposes the torn media to the intima (Krieger, 2015)
- Restenosis uncommon
- Allows for redilatation if needed as child grows, typically every 3-5 years (Beekman, 2008)
- See angiograms below for ciniaangiographic images of stent implantation
Actual Angiograms of Catheter Intervention of Native Coarctation with Stent Placement

- Covered stents
  - First covered stents (CP Covered stents) approved for use in CoA in 2016
  - Can be used to exclude an aneurysm or reduce bleeding after intimal tear (Krieger, 2015)

Actual Angiograms of Catheter Intervention of Native Coarctation with Placement of Covered Stent

- Outcomes
  - Mortality - rare beyond newborn period (Beekman, 2008)
    - Higher rate has been reported for angioplasty for recurrent post op CoA versus native CoA
  - Acute complications (Beekman, 2008)
- Femoral artery injury and thrombosis - common in infants younger than 12 months
- Femoral artery hemorrhage
- Cerebrovascular accident
  - Significant long term issues: See Section on Long Term Care below

**Long Term Care** (Vonder Muhull, 2016)
- Excellent prognosis for normal growth, development when CoA successfully repaired in childhood (Beekman, 2008)
- Lifelong care imperative to monitor for long term risks (Bhatt, 2015; Krieger, 2015)
  - Hypertension,
  - Re-coarctation,
  - Development of aneurysms
  - Premature cardiovascular complications
- CoA is a *Moderately complex* adult congenital heart condition (Adult Congenital Heart Association- Lifelong Care pamphlet)
  - Requires a minimum of an annual life time follow-up evaluation (Gurvitz, 2013)
  (See components of follow-up visit for adult care below.)
- Potential Complications & Risk
  - May occur after all forms of repairs (Kaemmerer, 2011)
  - **Residual CoA**
    - Presence of gradient in aorta after repair with the development of restenosis, gradient in aorta after an initially successful repair
    - 8% to 54% (Daniels, 2008)
    - Recoarctation
      - Suspected if upper and lower limb gradient of > 20 mm Hg
      - Measured noninvasively by blood pressure or directly by cardiac catheterization
      - May cause systemic hypertension, heart failure, left ventricular wall mass, coronary artery disease
      - Risk increases with younger age at time of repair
  - **Systemic arterial hypertension**
    - Present in 1/3 of patients
    - Increases over time even after technically successful intervention (Krieger, 2015)
    - Occurs at rest or during exercise (Krieger, 2015)
    - Target for BP therapy is < 140/90 (Bhatt, 2015)
    - More than 60% have hypertension 25 years after repair (Brown, 2013)
      - Can be related to re-coarctation. **If patients represent with hypertension after CoA repair, a residual obstruction must be ruled out
      - CoA patients have structural changes in the wall of vessels leading to stiffer arterial walls, reduced baroreceptor sensitivity, changes in renin-angiotensin system, impaired endothelial function
Higher risk of prevalence of hypertension with later repair (Bhatt, 2015)

Hypertension
- Can lead to early cardiovascular events
  - Third or fourth decade of life (Krieger, 2015)
  - Higher risk for myocardial infarction, cerebral vascular accidents, aortic dissection, LV systolic dysfunction, endocarditis (Krieger, 2015)

Coronary artery disease (CAD)
- Higher risk for premature onset atherosclerosis and death from coronary artery disease (Krieger, 2015)
- Important to monitor and control CAD risk factors
  - Hypertension
  - Hypercholesterolemia
  - Obesity
  - Smoking

Progressive valve disease, bicuspid aortic valve or mitral valve (Daniels, 2008)
- Bicuspid aortic valve can progress to stenosis (59-81%) or regurgitation (13-22%) (Sabet, 1999)
- Predictors of progressive valve dysfunction
  - Increasing age
  - Hypertension

Aortic aneurysm at the site of CoA, ascending or descending aorta
- Highest after prosthetic patch aortoplasty
- Increased risk of aortic rupture
- Recognition and early management essential to preventing a life threatening rupture
  - Imaging with MRI is the modality of choice
  - Can be managed with percutaneous covered stents

Brain aneurysm
- Dissection and intracranial hemorrhage
  - May be related to berry aneurysms in circle of Willis (Beekman, 2008)
- Higher risk of stroke

Long term concerns may be greatly affected by associated cardiac lesions
- Left shoulder elevation - seen in adults due to left lateral thoracotomy
- Left arm - decreased pulse/ BP if surgery used a left subclavian artery patch
- Sudden death (Daniels, 2008)

Bacterial endocarditis
- Antibiotic prophylaxis prior to dental procedures no longer required by American Heart Association, 2007
- Should seek additional information regarding status of other lesions

Long Term Follow-up Care in Adults with CoA repair
- Annual visits: Classified as moderately complex congenital heart disease
• Clinical evaluation: Monitor for re CoA (Krieger, 2015)
  ▪ Documentation of type of CoA repair is important
  ▪ Monitor blood pressures & pulses (Kaemmerer, 2011)
  ▪ Measure four extremity BP in arm in leg in lying flat at least yearly (Gurvitz, 2013)
    • Normal BP: Lower extremity BP will be higher than upper extremity BP by 10-20%
    • If lower extremity BP is lower than arm BP by > 10 mmHg then suspect a residual CoA or other form of peripheral arterial disease
    • If collateral vessels are present, the CoA gradient may not be high
  ▪ Pulses: Simultaneous palpation of right radial & femoral pulses: Suspect a CoA if the femoral pulse is weak or delayed in relation to the radial pulse
  ▪ Murmur – listen for posterior murmur
  ▪ Assessment NOTE: Monitor four extremities BP
    • If left subclavian artery used as part of repair, BP’s will be LOWER in the left arm (avoid BP measure & use of arterial line here)
    • If aberrant subclavian artery present – must consider use of left arm to obtain a BP which is proximal to CoA repair
    • May require ambulatory BP measures
  ▪ Electrocardiogram
  ▪ Transthoracic echo
  ▪ Cardiac magnetic resonance (MRI) or CT
    • Serial MRI surveillance aorta: potential aneurysms, pseudoaneysms, status of aortic repair, valves
    • Recommend – at least every five years (Gurvitz, 2013; Krieger, 2015)
    • With CoA stent present, less frequent use of MRI due to artifact in images
  ▪ Exercise test: Surveillance for exercise induced hypertension (Krieger, 2015)
  ▪ Monitor new or different type headache or chest pain
    • May be sign of possible cerebral aneurysm (Kaemmerer, 2011)
    • Report any chest pain or hemoptysis. Risk for aortic aneurysm formation, rupture (Vonder Muhll, 2016)
  ▪ Monitor closely for cardiovascular risk factors for CAD: control BP, cholesterol; avoid obesity & smoking (Kaemmerer, 2011)
    • Minimize additional risk for coronary artery disease
    • Treat modifiable risk factors (Krieger, 2015; Bhatt, 2015)
    • Aggressive medical treatment of residual hypertension – once a residual CoA is excluded (Krieger, 2015; Bhatt, 2015)
    • Encourage attainment of ideal body weight (BMI goal 18.5-25 kg/m2) (Bhatt, 2015)
• Encourage healthy eating, healthy life style, sodium restricted diet (Bhatt, 2015)
• Serial assessments lipid screening - LDL primary target for therapy, goal < 100 mg/dL (Bhatt, 2015)

• Education & Resources
  o Assess knowledge, review condition, life long care needs (Resources: American Heart Association (www.myamericanheart.org), Adult Congenital Heart Association (www.achaheart.org)
  o Pregnancy information website: http://www.heartdiseaseandpregnancy.com/
  o Annual education on risk for premature atherosclerotic heart disease risk factors, modifying the risk factors and self-care (Krieger, 2015)

Care during pregnancy (Refer to ACHD Guidelines on Pregnancy in Adults with CHD)
• Recommendations
  o Consultation: Adult congenital heart cardiologist before pregnancy
  o Collaborative, multidisciplinary care by adult congenital cardiology and perinatal team (Krieger, 2015)
• Patients at highest risk include:
  o Unrepaired CoA
  o Arterial hypertension
  o Residual CoA
  o Aneurysm at site of CoA repair (Kaemmerer, 2011)
• Risk of having child with a heart defect 3-10% (ACHA)

References:


Revised July 2016
JA Nieves, A Green