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

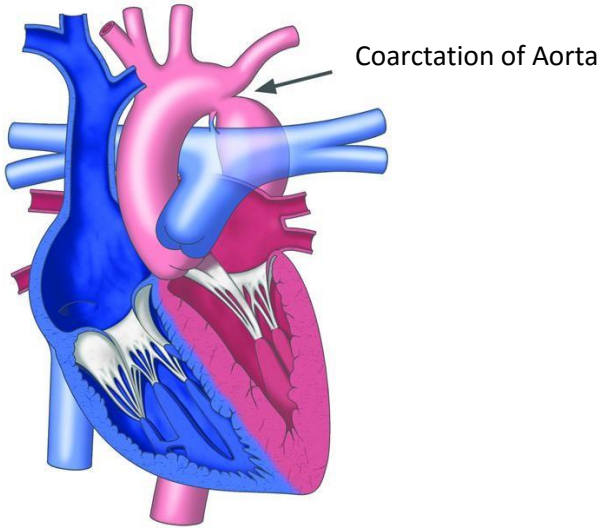
Amanda Green, MSN, ARNP, FNP-C
Nurse Practitioner, Cardiac Catheterization
Nicklaus Children's Hospital
Miami, Florida

Embryology

- Affects 5% to 8% of all newborns with congenital heart disease (Krieger, 2015)
- Occurs during the 6th to 8th week of gestation
    - 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).
    - 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)
    - Long hypoplastic segment- a tubular hypoplasia involving the aortic arch or the aorta distal to the orgin 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

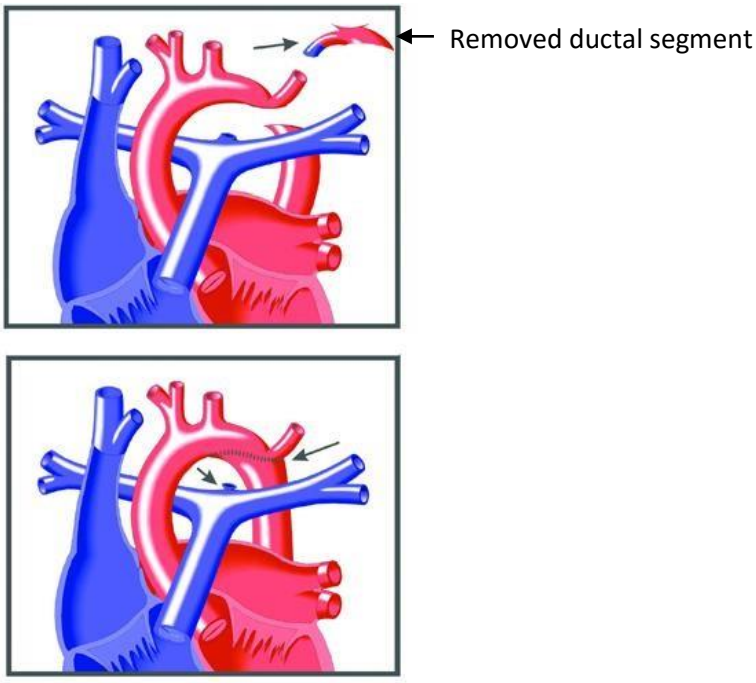
- Cardinal features (Krieger, 2015; Kaemmerer, 2011)
  - Upper body arterial hypertension
  - Weak, absent, and/or delayed femoral pulses
  - Decrease in blood pressure in lower extremities
  - Palpable collateral arteries over the medial aspect of the scapulae, the lateral chest wall, and between the 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:
                - Shock & heart failure
                - Metabolic disturbances
                - Hypothermia
                - Hypoglycemia
        - Results in: (Beekman, 2008)
            - Lower body
            - Renal hypoperfusion with renal failure
            - Necrotizing enterocolitis (NEC)
- Child or adolescent
    - 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
    - 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
    - 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

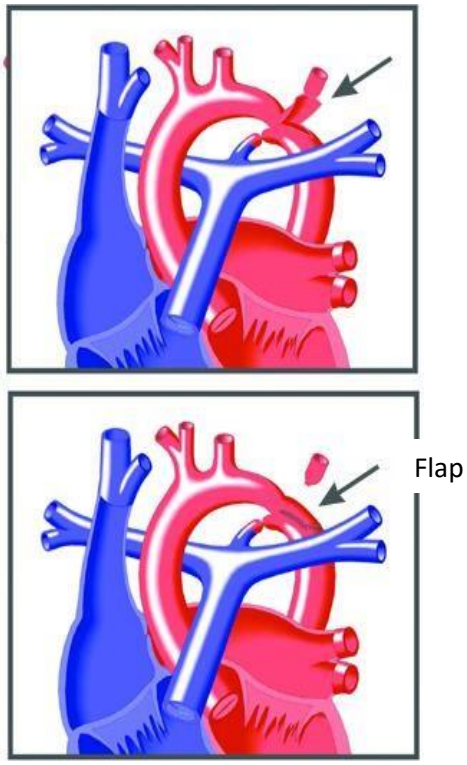
- Diagnosis:
    - Most often via clinical exam, echocardiogram, and chest x-ray, MRI or CT
    - Diagnostic cardiac catheterization only if anatomy and hemodynamics, associated lesions are more complex, and additional clinical questions are present (Beekman, 2008)
- Treatment & Timing:
    - Individualized to lesion, associated conditions
    - 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
                - 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
    - Child, adolescent
        - Repair at 2 to 3 years of age, or upon diagnosis
    - 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
    - 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)



End-to-End Anastomosis with removal of Ductal Segment
Illustrations reprinted from PedHeart Resource. www.HeartPassport.com.

    - 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

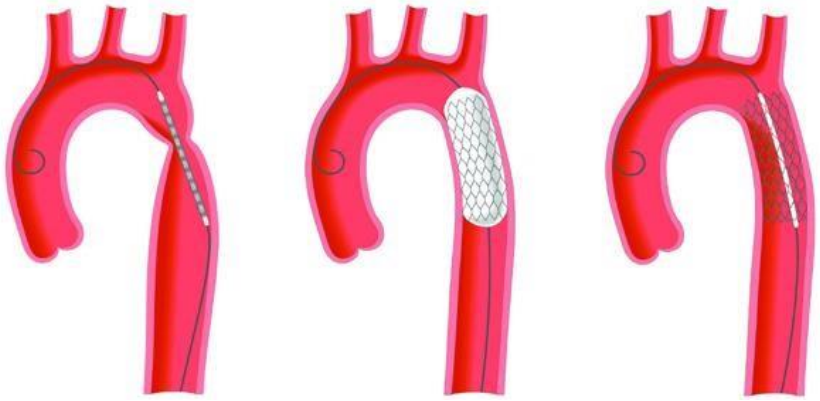


Coarctation Repair with Left Subclavian Flap
Illustrations reprinted from PedHeart Resource. www.HeartPassport.com.

- 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

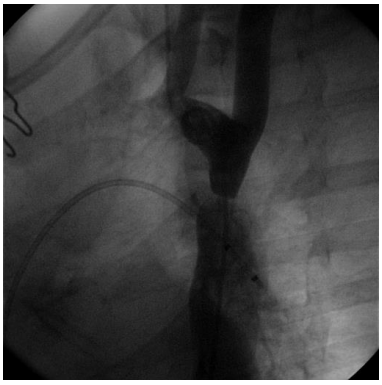
- Cardiac Catheterization: Interventional, Balloon angioplasty, potential stent
    - Balloon angioplasty
        - Began 1982
        - Widely accepted for treating re-coarctaton
        - Enlarges CoA lumen
            - Produces linear intimal and medial tears at the CoA site
            - Artery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



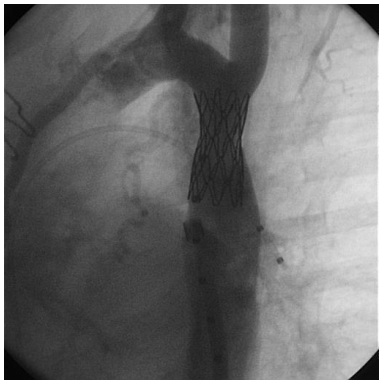
Balloon Angioplasty with Implantation of Stent
Illustrations reprinted from PedHeart Resource. www.HeartPassport.com.

- 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 ciniangiographic images of stent implantation

Initial Angiogram demonstrating severe native coarctation of the aorta with near interruption.



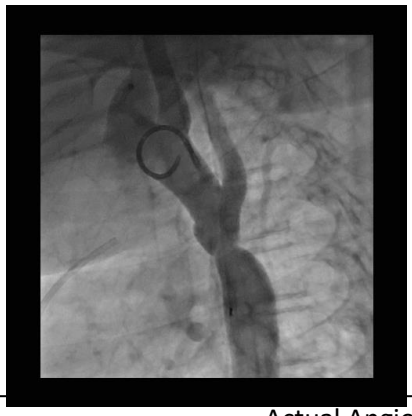
Final angiogram post CP bare metal stent angioplasty in the same patient.



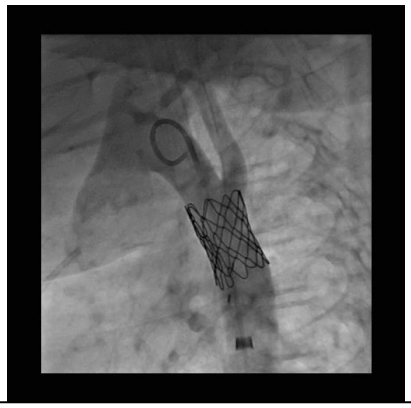
Actual Angiograms of Catheter Intervention
of Native Coarctation with Stent Placement

- 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)

Initial Angiogram demonstrating coarctation of the aorta in a 9 year old with Turners Syndrome and Shones complex.



Final angiogram post CP covered stent angioplasty in the same patient.



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 Muhull, 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
    - Assess knowledge, review condition, life long care needs (Resources: American Heart Association (www.myamericanheart.org), Adult Congenital Heart Association (www.achaheart.org)
    - Pregnancy information website: http://www.heartdiseaseandpregnancy.com/
    - ACHA Q and A: Coarctation of the Aorta; Adult Congenital Heart Association. http://www.achaheart.org/Portals/0/pdf/Library%20Education/Coarctation2014.pdf Accessed May 15, 2016.
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    - 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
    - Consultation: Adult congenital heart cardiologist *before* pregnancy
    - Collaborative, multidisciplinary care by adult congenital cardiology and perinatal team (Krieger, 2015)
- Patients at highest risk include:
    - Unrepaired CoA
    - Arterial hypertension
    - Residual CoA
    - Aneurysm at site of CoA repair (Kaemmerer, 2011)
- Risk of having child with a heart defect 3-10% (ACHA)

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JA Nieves, A Green