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### Coarctation of the Aorta

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# Coarctation of the Aorta

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## Introduction

Coarctation of the Aorta (CoA) is one of the most common congenital heart diseases, accounting for 6%-8% of all congenital cardiac diagnoses (Ganigara et al., 2019). CoA is a constriction of the aortic isthmus, most often in the area between the left subclavian artery and the ductus arteriosus (Joshi et al., 2017). Intervention for CoA is based on the severity of the constriction, and includes surgery and balloon angioplasty with or without balloon placement (Fox et al., 2019).

Knowledge of the pathophysiological process of CoA is vital for all practice levels of nurses. Although most cases of CoA are diagnosed in early childhood and are managed by pediatric practitioners, some cases do present in adults.

## Signs and Symptoms

Presentation of CoA is determined by the degree of coarctation of the aorta. In patients with severe CoA, the following symptoms typically present within the first week of life when the patent ductus arteriosus (PDA) restricts or closes.

- Tachypnea
- Blood pressure gradient between upper and lower extremities
- Low cardiac output
- Metabolic acidosis
- Absent or decreased femoral pulses
- Differential cyanosis

In patients with less severe coarctation or presence of collateral vessels, patients may not become symptomatic for weeks, months, or years after closure of the PDA (Joshi et al., 2016). These patients may present with symptoms previously described in addition to the following:

- Failure to thrive
- Left ventricular hypertrophy
- Cardiomegaly
- Hypertension

## Clinical Presentation

A six day old male presents to the Emergency Department for a one day history of decreased enteral intake, fussiness, and tachypnea. The patient was born vaginally and had an unremarkable hospital course before being discharged four days ago. The parents report that the child's last wet diaper was six hours ago.

Upon physical assessment, the RN notes that the patient has a loud murmur, mottling of the skin, decreased lower extremity pulses, subcostal retractions, and abdominal muscle usage.

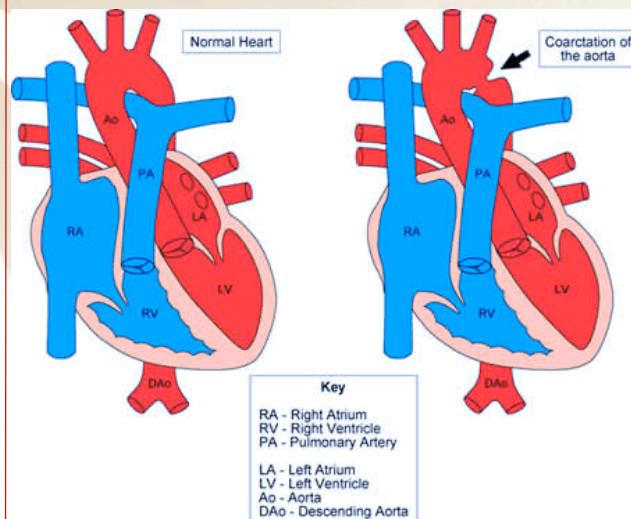
Vital signs taken on the right arm are as follows:

- HR 174
- BP 58/37 (43)
- SpO2 93%
- RR 74
- Temp 97.8 F.
- A lower extremity blood pressure is also obtained: 43/22 (29).

Findings are reported to the Advanced Practice Nurse, who orders a stat capillary blood gas, which resulted

- pH 7.27
- pCO2 38
- HCO3 16
- pO2 30

The patient is placed on nasal cannula 4 liters, an IV is placed, and alprostadil is started. The patient is admitted to the Cardiac ICU where an ECHO indicates Coarctation of the Aorta with a restricted ductus arteriosus.



Nationwide Children's Hospital. What is Coarctation of the Aorta? [Digital image]. Retrieved July 31, 2020, from <https://www.nationwidechildrens.org/conditions/coarctation-of-the-aorta>

## Underlying Pathophysiology

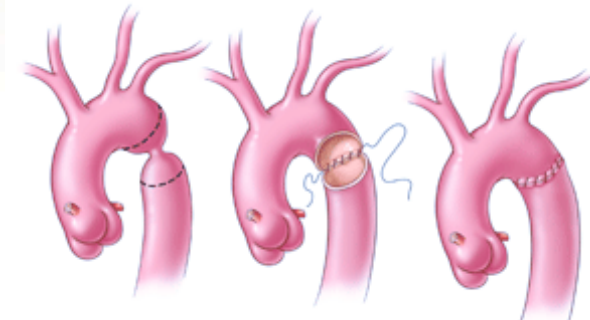
- There are two commonly accepted hypothesis for the etiology of CoA (Ganigara et al., 2019)
  - Hemodynamic Theory: Decreased flow through the aortic isthmus during fetal life lead to isthmus hypoplasia and CoA
  - Ectopic Ductal Tissue Theory: Extension of ductal tissue into the aorta causes the aortic isthmus to contract and sclerose in the same manner as the PDA
- Classic Neonatal Presentation
  - At birth, most often a patient with CoA has an unremarkable physical assessment
  - As the coarctation tissue begins to constrict, deoxygenated blood from the PDA will shunt into the descending aorta, causing differences in oxygen saturation between the upper and lower extremities
  - After pulmonary vascular resistance drops and the PDA closes, there will be inadequate blood flow to organs distal to the coarctation causing (Fox et al., 2019):
    - lower body ischemia
    - electrolyte imbalance
    - decreased renal perfusion
  - Eventually severe acidosis from ischemia will lead to circulatory collapse CoA (Ganigara et al., 2019)
- Later Presentation
  - CoA can go undetected in some patients. Due to the coarctation, these patients will have:
    - high left ventricular afterload which can lead to:
      - left ventricular hypertrophy
      - pulmonary edema
    - aortic isthmus constriction which can lead to:
      - arterial collaterals
      - upper limb hypertension
      - ischemic or hypertensive organ damage

## Implications for Nursing Care

- Pre-operative
  - Apnea is a common side effect of alprostadil. Respiratory support such as positive pressure ventilation may be needed (Joshi et al., 2017)
  - Four extremity blood pressures must be taken frequently to ensure adequate blood pressure to the lower extremities
  - Pre-ductal and post-ductal oxygen saturation monitoring to ensure adequate oxygenation to the lower extremities (Fox et al., 2019)
- Post-operative
  - Hypertension is common after CoA repair. The nurse must closely monitor the patient for appropriate blood pressure and be prepared to administer anti-hypertensive medications (Fox et al., 2019)
  - Four extremity blood pressures must be taken frequently to assess for the presence of re-coarctation (McCance & Huether, 2019)
- Family Centered Care
  - The nurse must encourage patient family interactions before and after surgery to promote normal bonding
  - Education about CoA, surgical repair, side effects, and long term implications must be provided

## Treatment

- Alprostadil
  - A prostaglandin used in patients with CoA to maintain ductal patency prior to repair
  - Maintain bloods flow distal to the site of coarctation
  - May also be initiated in patients whose PDA has already closed to relax the ductal tissue at the sit of the coarctation (Fox et al., 2019)
  - Common side effects include: apnea, fever, and hypotension (Munoz, 2014)
- End to end anastomosis of the aorta
  - The aorta is mobilized, the coarctation is isolated, and the area is resected and then reanastomosed (Fox et al., 2019)
  - Surgical method of choice for neonates with coarctation of the aorta (Joshi et al., 2017)
- Percutaneous Catheter Interventions
  - Balloon angioplasty with or without stent placement is the treatment of choice for older children or adults with discrete coarctation (Fox et al., 2019)
  - Balloon angioplasty is the preferred intervention for recurrent coarctation in children (Joshi et al., 2017)



Cleveland Clinic. (2019, May 14). End-to-end anastomosis [Digital image]. Retrieved from <https://mv.clevelandclinic.org/health/diseases/16876-aortic-coarctation/management-and-treatment>

## Significance

- CoA is the most commonly missed ductal dependent lesion on neonatal exam (Beattie et al., 2017)
- The majority of neonatal CoA cases are diagnosed late and many infants only present to the hospital when they are critically ill (Sorenson et al., 2020)
- Recurrent aortic arch obstruction intervention is indicated in up to 20% of surgically repaired neonates (Adamson et al., 2017)
- Patients with CoA have a higher likelihood of coronary artery disease, aortic aneurysm, and brain aneurysm (Nieve & Green, 2016)
- 30% of patients with CoA presenting in childhood have associated congenital heart defects (Joshi et al., 2017)
- Patients with CoA have unique needs and can be medically fragile. It is essential for practitioners to have an understanding of the signs, interventions, and comorbidities for this disease to ensure best patient outcomes.

## Conclusions

- CoA is a ductal dependent heart defect that results in decreased blood flow to areas of the body distal to the point of constriction
- Priorities for nursing care ensure appropriate blood flow and oxygenation to both pre-ductal and post-ductal areas of the body
- Repair of CoA may be performed surgically or using balloon angioplasty
- Patients with CoA have increased risk of developing other vascular lesions
- CoA is often goes undetected until patients face circulatory collapse. More accurate and valid forms of diagnosis are warranted.

## References



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