GUIDELINE

Cardiac: Coarctation of the Aorta (CoA) and Interrupted Aortic Arch (IAA) – Pre-operative Management

Scope (Staff):	Nursing and Medical Staff
Scope (Area):	NICU KEMH, NICU PCH, NETS WA

Child Safe Organisation Statement of Commitment

CAHS commits to being a child safe organisation by applying the National Principles for Child Safe Organisations. This is a commitment to a strong culture supported by robust policies and procedures to reduce the likelihood of harm to children and young people.

This document should be read in conjunction with this disclaimer

Contents

Aim

This guideline outlines the identification and pre-operative management of coarctation of the aorta and interrupted aortic arch in the neonatal population.

Risk

Failure to follow this guideline may lead to adverse outcomes for the infant.

Definitions

CoA is defined as a constriction or narrowing of the aorta which is variable in severity. It becomes significant when there is a pressure gradient of > 20 mmHg across the aortic narrowing. More than 60% of infants have an associated bicuspid valve (of note, only a small proportion with a bicuspid aortic valve have a CoA) and in about 50% there are other associated cardiac lesions such as VSD, ASD, PDA, HLHS, TGA, mitral valve abnormalities, aortic stenosis. There is also higher incidence with non-cardiac conditions such as congenital diaphragmatic hernia.

Background

Coarctation of the Aorta (CoA) accounts for 6-12% of all congenital heart disease and has an incidence of approx. 1 in every 1200-1300. It is more common in males in a ratio of 2:1. Interrupted aortic arch (IAA) is rare, accounting for only 1% of critically ill neonates with congenital heart disease. The clinical presentation is similar to that of CoA, so is discussed here. CoA usually occurs in the thoracic aorta around the ductus, but rarely, it occurs in the abdominal aorta. It results from thickening of the media of the aortic wall that forms a ridge on the inner surface of the aorta. There is often post-stenotic dilatation of the descending aorta, but if the stenosis is severe, there may be hypoplasia of the proximal descending aorta, often referred to as long segment coarctation. There may also be narrowing of the transverse and isthmic portion of the aortic arch known as a hypoplastic arch.

CoA is frequently a component of several chromosomal abnormality syndromes. The exact genetic and molecular abnormality of this defect is unknown. It is relatively common in trisomy 13 and 18, Turner's (Monosomy X), Williams and Kabuki syndromes. 15-20% of infants with Turner's Syndrome have CoA. IAA is associated with 22q1.1 deletion.

Physiology of the Lesion

Narrowing in the aorta leads to decreased pressure or hypotension distal to the obstruction causing decreased perfusion of major organs and muscle. Decreased perfusion of the kidneys causes them to secrete renin which causes arterial vasoconstriction in an attempt to raise the distal pressure and so causing hypertension proximal to the obstruction. The left ventricle is subjected to increased pressure (afterload) which may lead to significant hypertrophy. With an acute increase the afterload, the left ventricle may not be able to compensate, and clinical heart failure may occur. If this occurs, blood pressure may be decreased both proximally and distally to the obstruction.

In the presence of complete or near complete juxtaductal obstruction and a patent ductus arteriosus in the presence of raised PVR blood can still flow to the distal aorta

(as is the case with neonates). This occurs by R→L shunting across the ductus to supply the abdominal organs and lower extremities. Therefore, blood flowing to the distal aorta will be desaturated.

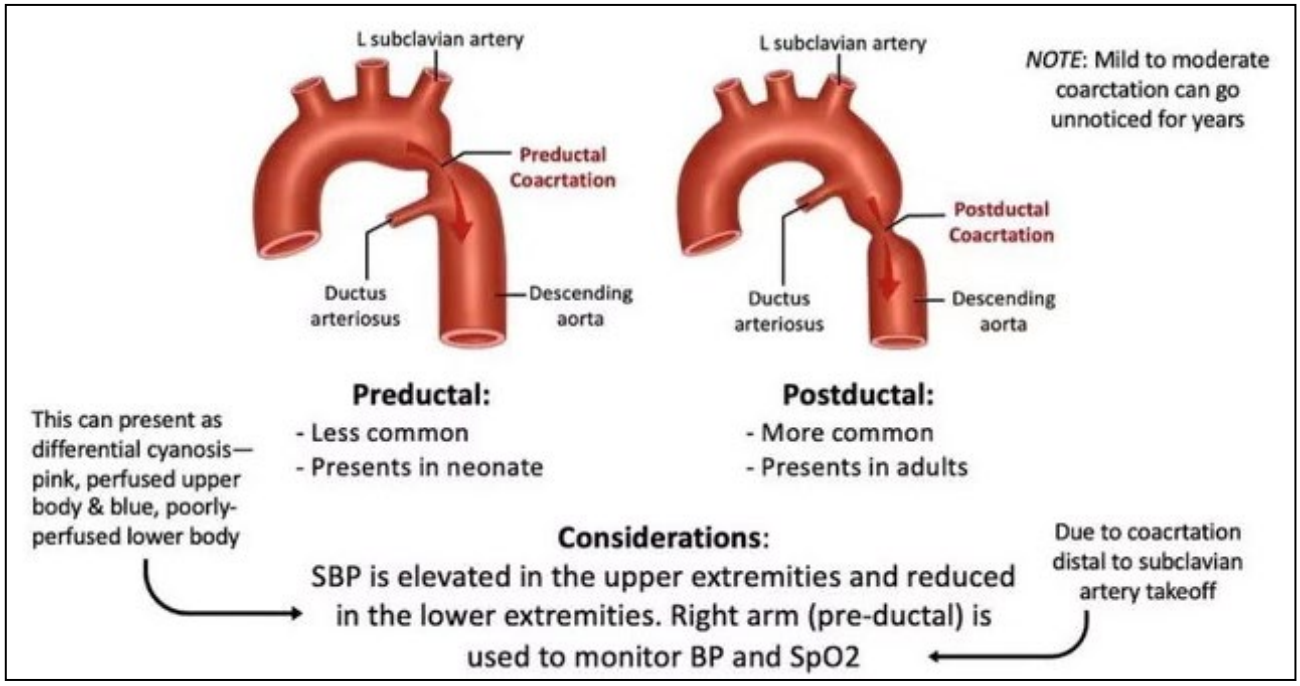
If the obstruction is long-standing (those that present out of the neonatal period), collateral vessels develop from the proximal to the obstruction.

Classification of CoA

There are several classifications of CoA, the most useful is to divide the condition into pre-and post-ductal CoA, which classifies the blood flow of the anomaly.

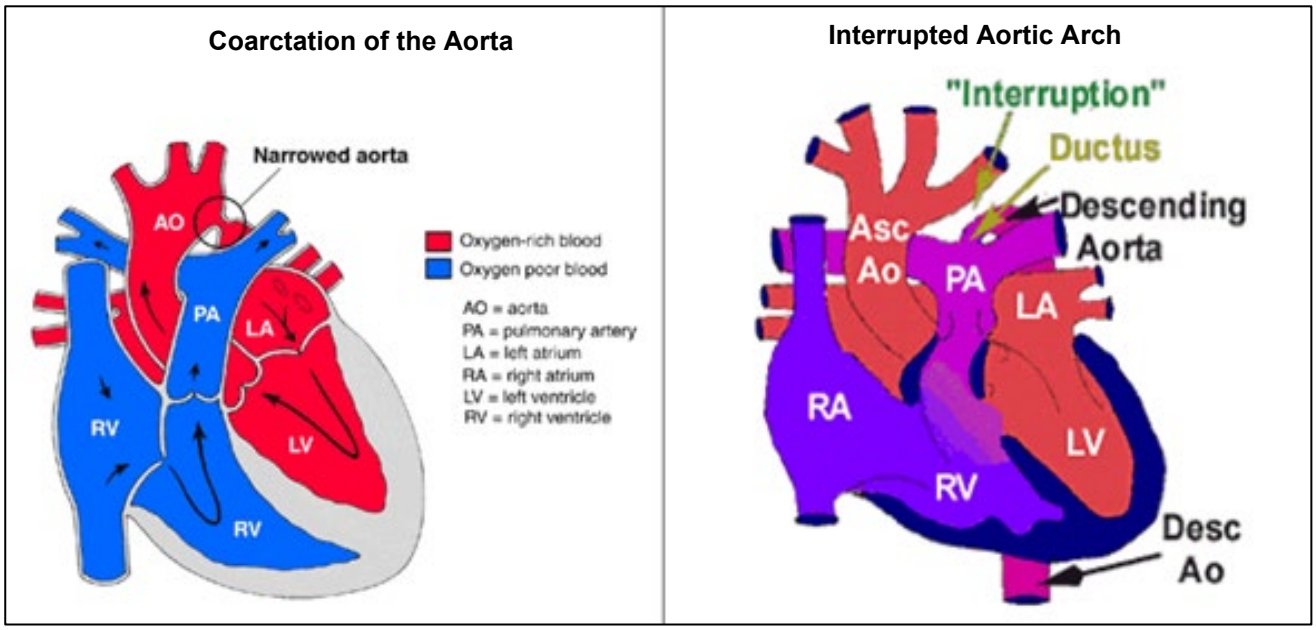
Pre-ductal - The CoA is found proximal to the ductus arteriosus. It is frequently associated with other anomalies. As the ductus supplies the descending aorta in the fetal period, there are not many aortopulmonary collaterals to support the descending aorta. Hence, this type presents early in the neonatal period as soon as the ductus starts constricting.

Post-ductal (Juxtaductal) - This is the most common form and is found distal to the ductus arteriosus. As the coarctation is present distal to the ductus arteriosus, there are aorto-pulmonary collaterals formed in the fetal life and can support the descending aorta even when the ductus starts constricting in the post-natal life. This type usually presents later in the infantile or even the adult age group depending upon the collateral supply.



Another way to classify CoA is:

- Isolated CoA.
- CoA with VSD.
- CoA with complicated intracardiac anomaly.



Clinical Presentation

There are 3 distinct presentations in neonates:

1. **Antenatally** - diagnosis of coarctation of aorta in the fetal period is very challenging and in the best of the centres is only around 55% (roughly 1 out of 2 cases are missed on routine fetal cardiac scans).
2. **Routine newborn check** with pre and post ductal saturation monitoring. See Pulse Oximetry Screening to Detect Critical Congenital Heart Disease (CHD), the neonate is usually well, without any acidosis, cardiac failure or end-organ compromise. Occasionally, a murmur of a PDA or VSD is heard at the physical check which leads to further investigations, but usually, symptoms first present when the ductus begins to close or when there is associated LV failure
3. **Collapse in the first couple of weeks of life** when the DA closes. When an unrecognised CoA presents in the neonatal period, it is most often severe. They are usually asymptomatic at birth and become symptomatic as the ductus arteriosus closes at 4-10 days age. The neonate has been receiving desaturated blood to the lower extremities until this time, but this provides adequate perfusion to the abdominal organs and lower extremities.

    - The initial symptoms may be decreased/poor feeding regulation with shortness of breath and sweating or vomiting due to poor gut perfusion.

- As the duct shuts, shock ensues:
  - Grey/mottled appearance with lethargic/decreased level consciousness.
  - Prolonged capillary refill in lower extremities.
  - Absent femoral pulses (full pulses in right arm).
  - Differential blood pressure between lower extremities and right arm.
  - Tachypnoea/recessions.
  - Differential in pre- and post-ductal SaO2.
  - Enlarged liver.
  - Oliguria/anuria.
  - Metabolic acidosis/high lactate (often severe).

Management of CoA/IAA

1. Initial Management at Birth if Stable with Suspected CoA

- Infants diagnosed antenatally are unlikely to be unwell at birth, so follow usual postnatal resuscitative measures.
- Infants suspected of CoA following routine pre and post ductal saturation monitoring at birth are unlikely to be unwell. If born at KEMH, admit to SCN3, assess and contact cardiologist early. The Cardiologist will decide when and what dose to commence Alprostadil (PGE1).
- If referred from a peripheral hospital (i.e. via NETS referral), involve cardiology in referral conversation. Consider commencing PGE1. PGE1 is not only helpful in opening up the ductus arteriosus but can sometimes assist with opening up of the coarctation segment itself. Make arrangements for urgent transfer to Ward 3B, PCH by NETS WA.

2. Initial Management If Presenting Collapsed/Shocked

Involve cardiology with management plan early. Perform the following investigations and imaging. See pre-operative management.

Airway/Breathing

- Strongly consider mechanical ventilation to decrease metabolic demand (and needing PGE1 infusion). Use pressures/FiO2 to keep normocarbia. Avoid hypocarbia which can lead to drop in pulmonary artery pressures and reduce the R→L flow in the ductus arteriosus to support the flow in descending aorta. Monitor SpO2 in right hand, maintain normoxemia (94-97%).
- Use small amount of sedation for intubation and as ongoing infusion, so as not to compromise cardiac function further. Muscle relaxant agents should be avoided as can cause hemodynamic instability with low blood pressures.

Circulation

- Establish good IV access (initial peripheral IV or UVC). Insert a second venous access if prostaglandins are required, as they need a dedicated line for infusion.
- Give normal saline fluid boluses in 10 mL/kg aliquots, maximum of 2. If requiring more than this, discuss with consultant as may make cardiac failure worse
- Commence PGE1/Alprostadil as soon as possible at 50ng/kg/min. Femoral pulses should be assessed until they can be palpated once PGE1 infusion has commenced, then reassess 4 hourly or if there are clinical concerns.
- Monitor the blood pressure closely. Pre-ductal non-invasive (cuff) blood pressure monitoring should be done regularly from the right upper limb. Monitoring from the lower limb or invasive blood pressures from descending aorta will suggest low blood pressures and should not be used as a guide to start inotropes.
- The left ventricle may need support with inotropes. However, this needs to be discussed with the on-call neonatal and cardiology consultants about the type of inotrope to be used. Caution to be exercised for inotropes that cause exclusive systemic vasoconstriction as they can potentially worsen the cardiac function by increasing the afterload.
- Arterial lines are helpful but do not delay treatment for this. A UAC is helpful and adequate and gives a post-ductal BP reading which is useful for assessing post-ductal perfusion. Do not have multiple attempts for peripheral arterial cannulation as that might reduce the chances for subsequent cannulation in theatre by cardiac anaesthetists. The right upper limb arteries should be avoided as a line is required here by the anaesthetist for theatre

Consider other causes of clinical deterioration/collapse

- Sepsis/Meningitis
- Shock
- Bleeding such as trauma/NAI, Vit K deficiency
- Intra-abdominal pathology such as volvulus
- Other cardiac causes
- Inborn errors of metabolism
- Endocrine causes (e.g. Congenital Adrenal Hyperplasia)

Investigations

Blood Gas	If presenting in shock, may show severe metabolic/ lactic acidosis which may be partially compensated with a low pCO2.
CRP/ Blood Culture	Check for evidence of sepsis
Blood Group and Hold / FBC	May need PRBC transfusion if in cardiac failure (worsened by low Hb)
Coagulation profiles	May be deranged (particularly INR) if period of hypoperfusion/ shock.
U+E /Ca/Mg /LFTs	Creatinine may be elevated. Any infant in shock or when 22q11 deletion possible (e.g. IAA). Ionised Ca on gas most helpful. Transaminases may be elevated.
Chromosomal analysis or microarray	If dysmorphic features/other anomalies and send for 22q11 deletion if IAA.
CXR	May show cardiomegaly and ↑ pulmonary vascular markings secondary to heart failure. Check position of tubes/lines.
Cardiac Echo	The prominent features are a narrowed aortic segment at the site of the CoA and post-stenotic dilatation of the descending aorta. Doppler is used across the defect to determine the pressure gradient. Occasionally the CoA is not visualised as the PGE1 has opened up the CoA itself. Echo also determines other associated defects, whether there is adequate flow through the PDA and the function of the left ventricle.
Cranial USS	Perform prior to theatre for those who present unwell. If need for interstate transfer, consider doing MRI brain if abnormal cranial USS or dysmorphic features or known genetic anomalies.
Renal USS	If there are other anomalies/chromosomal abnormality suspected

Ongoing Preoperative Management

Operative repair is carried out once the acidosis and multiorgan failure have settled.

Circulation

- Continue PGE1/Alprostadil until theatre. The dose may be decreased on the advice of the cardiologist.
- Ongoing inotrope support as needed.
- Monitor lactate levels closely:
  - Hourly initially until patient stabilising and metabolic acidosis improving.
  - 2 hourly until metabolic acidosis/lactate normalised.
  - 4 hourly unless clinical deterioration.

- If lactate it is not improving or starts rising, femoral's should be felt and consultant +/- cardiologist informed.

Respiratory

- Most patients having presented collapsed/ in shock to remain mechanically ventilated to maintain normocarbia. Hypocarbia should be avoided as they can lead to drop in pulmonary artery pressures and reduce the R→ L shunting through the ductus arteriosus.
- Normal SIPPV+VG (KEMH) or PC-AC+VG (PCH) with normal term neonatal settings for PEEP and pressures should be used. A higher PEEP may be required if there are signs of pulmonary oedema and may encourage the desired R > L shunt.
- The minimum FiO2 to keep the pre-ductal O2 sats 94-97% should be used.

Fluids/Nutrition

- Fluid restrict, total volume will depend upon postnatal age. In infants > 4 days old, 80 mL/kg/day 10% Dextrose + 0.22% Saline should be used (with potassium added as required) initially and liberalised depending upon the clinical status.
- Further saline boluses are sometimes required but should be used judiciously and only 5mL/kg at a time with reassessment prior to further.
- Monitor electrolytes closely, particularly K and Ca and correct accordingly.
- Keep NBM prior to theatre as there is a very high risk of NEC due to prolonged hypoperfusion of the gut and the following reperfusion injury. Start TPN.
- Monitor urine output hourly (should be >1mL/kg/hr). A urinary catheter may be required. Sometimes Frusemide is necessary especially in heart failure

Sepsis

As sepsis is common in neonates, most will be given at least 48 hours IV antibiotics until sepsis can be excluded.

Haematology

Correct coagulopathy/ low platelet count as necessary.

Possible Pre-Operative Complications

- Renal impairment/ failure.
- Cardiac failure.
- Coagulopathy.
- Multiorgan failure.
- Necrotising enterocolitis.

Related CAHS internal policies, procedures and guidelines

- Cardiac: Coarctation of the Aorta (CoA) Management Following Surgical Closure
- Cardiac: General Complications Management Following Surgery
- Cardiac: Routine Post-Operative Care
- Pulse Oximetry Screening to Detect Critical Congenital Heart Disease (CHD)
- King Edward Memorial Hospital - Neonatal Medication Protocols (health.wa.gov.au)

References and related external legislation, policies, and guidelines

1. Raza, S., Aggarwal, S., Jenkins, P., Kharabish, A., Anwer, S., Cullington, D., Jones, J., Dua, J., Papaioannou, V., Ashrafi, R., & Moharem-Elgamal, S. (2023). Coarctation of the Aorta: Diagnosis and Management. Diagnostics (Basel, Switzerland), 13(13), 2189. https://doi.org/10.3390/diagnostics13132189

2. Sun HY, Proudfoot JA, McCandless RT. Prenatal detection of critical cardiac outflow tract anomalies remains suboptimal despite revised revised obstetrical imaging guidelines. Congenit Heart Dis. 2018;13:748–756

3. Arya, B., & Maskatia, S. A. (2022). Coarctation of the aorta: Prenatal assessment, postnatal management and neonatal outcomes. Seminars in perinatology, 46(4), 151584. https://doi.org/10.1016/j.semperi.2022.151584

4. Nichols, D.G.; Ungerleider, R.M.; Spevak, P.J.; Greeley, W.J.; Cameron, D.E.; Lappe, D.G.; Wetzel, R.C., Critical heart disease in infants and children. Second ed.; Mosby: USA, 2006.

5. Artman, M.; Mahony, L.; Teitel, D.F., Neonatal cardiology. Second ed.; USA, 2011.

6. Horrox, F., Manual of neonatal and paediatric heart disease. First ed.; Whurr Publishers: Gateshead, Tyne and Wear, UK, 2002.

7. Koenig, P.; Hijazi, Z.M.; Zimmerman, F., Essential pediatric cardiology. First ed.; McGraw-Hill Companies, Inc: USA, 2004

This document can be made available in alternative formats on request.

Document Owner:	Neonatology		
Reviewer / Team:	Neonatal Coordinating Group		
Date First Issued:	April 2014	Last Reviewed:	August 2024
Amendment Dates:		Next Review Date:	August 2027
Approved by:	Neonatal Coordinating Group	Date:	27th August 2024
Endorsed by:	Neonatal Coordinating Group	Date:	
Standards Applicable:	NSQHS Standards:  Child Safe Standards: 1,10		

Printed or personally saved electronic copies of this document are considered uncontrolled

Healthy kids, healthy communities

Compassion | Excellence | Collaboration | Accountability | Equity | Respect

Neonatology | Community Health | Mental Health | Perth Children's Hospital