the children's hospital at Westmead

COARCTATION REPAIR IN NEONATES- GCNC - CHW

PRACTICE GUIDELINE ©

DOCUMENT SUMMARY/KEY POINTS

- Coarctation of the aorta is a congenital heart defect where the aorta is narrowed obstructing left heart outflow.
- The diagnosis may be suspected antenatally or following presentation in cardiogenic shock.
- Echocardiography confirms the diagnosis and associated anatomy.
- Pre-operative management consists of providing appropriate respiratory and circulatory support, maintaining or reopening the duct with prostaglandin E1and carefully managing fluids and electrolytes. Enteral feeding is deferred until flow into the aorta is restored and is preferably with expressed human milk or Pasteurised donor human milk (PDHM).
- Post-operative management includes support with invasive mechanical ventilation to allow adequate analgesia post-thoracotomy, control of systemic hypertension, fluid and electrolyte balance and vigilance for immediate post-operative complications.
- Potential complications include necrotising enterocolitis and vocal cord dysfunction.

Approved by:	SCHN Policy, Procedure and Guideline Committee	
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NSW GOVERNMENT

CHANGE SUMMARY

- Document due for mandatory review. Changes made throughout. Recommend to real the entire policy.

READ ACKNOWLEDGEMENT

- All clinical staff working in Grace Centre for Newborn Intensive Care.
- Read Acknowledge Only – [all staff emailed the notice of the new document and expected to read and be aware of practices and changes]

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Background

Coarctation of the aorta is a congenital heart defect with an incidence of 3-5 per 10,000 live births in Australia with a slight male preponderance [1, 2]. It consists of a narrowing of the aorta which impairs blood flow. The narrowing most commonly occurs peri-ductally in the descending aortic arch. Coarctation can occur in isolation or as a component of more complex structural heart disease. This guideline deals with the more common situation of discrete Coarctation with or without an intact ventricular septum. The principles outlined in this document may not apply in the presence of a more complex lesion.

Presentation

The clinical features of coarctation are dependent on the site and degree of obstruction. In the Grace Centre for Newborn Intensive Care, suspected coarctation usually presents via one of three pathways. The first two arise as a result of antenatal suspicion with admission and evaluation prior to any development of left heart obstruction. The third pathway usually occurs when there is no antenatal suspicion and presentation is a result of obstruction following postnatal ductal closure.

Suspected Coarctation where ductal patency is not maintained with prostaglandin

In this situation, the presence of Coarctation is suspected by the fetal cardiologist on antenatal ultrasound, however, there are no clinical or laboratory signs of impairment of distal blood flow and the cardiologist recommends no intravenous prostaglandin E_1 (PGE_1) at the time of delivery but a period of close observation. Echocardiographic definitive diagnosis of Coarctation is difficult in the presence of a patent ductus although there are some features that can heighten suspicion [3] [4]. Here, a period of observation occurs whilst the ductus closes in order to ensure that Coarctation does not evolve.

Suspected Coarctation where ductal patency is maintained with prostaglandin

This presentation is similar to the previous except that ductal patency is maintained with intravenous prostaglandin at the time of admission. Either the diagnosis is confirmed on echocardiography and ductal patency is maintained pending surgical repair or following postnatal echocardiography, the index of suspicion is low and prostaglandin is discontinued and the baby closely observed whilst the ductus closes.

Suspected Coarctation with significant features of obstruction

The neonate with critical Coarctation of the aorta usually presents with congestive cardiac failure and loss of systemic blood flow when the ductus closes – mostly around 3-10 days of postnatal age. The location and degree of constriction will determine the severity and timing of the presentation. Less critical Coarctation may present outside of the neonatal period [5].

The features are those of congestive heart failure with diminished distal blood flow and include respiratory distress with weak or absent femoral pulses accompanied by metabolic acidosis and an elevated lactate. There may be associated increase in the brachial pulses along with renal and hepatic impairment. Since this presentation is similar to other causes of shock, including sepsis, an active praecordium, marked hepatomegaly and cardiomegaly with pulmonary plethora on CXR are valuable clinical differentiators [5]).

Blood pressure differential between the right upper limb and the lower limbs is not useful in the neonatal period although it is often thought to be an important finding [6]. In the early acute presentation with shock, all pulses may be difficult to appreciate and their absence should not exclude the possibility of left heart obstruction being present.

Pre-operative management

The clinical or antenatal diagnosis of Coarctation is confirmed on arrival into the intensive care with echocardiogram by the paediatric cardiologist. The Cardiology team confirms the anatomy of Coarctation and any associated lesions such as ventricular septal defect and mitral valve or aortic valve anomalies. Occasionally, a CT angiogram is required in addition to echocardiography to appreciate the exact anatomy present.

Once the diagnosis of Coarctation requiring surgical repair is established, the pre-operative management consists of continuous monitoring of clinical signs, maintenance of intravenous access to ensure PGE_1 delivery and ductal patency, respiratory and cardiovascular support as required, and completion of preoperative surgical screening. Urinary catheter may be considered useful depending on illness severity preoperatively.

Preoperative investigations

Infants with suspected or proven Coarctation should have a chest X-ray, a full blood count, LFTs, coagulation studies, group and hold (pending crossmatch), serum assessment of electrolytes, urea, creatinine, calcium, magnesium and phosphate. Fish for 22q11 and a CGH microarray is required particularly in female infants, because of the association with Turner's syndrome [8]. Newborn screening should be collected pre-operatively in case blood transfusion is required.

Monitoring

Infants with established Coarctation are routinely admitted to intensive care and managed on an open care radiant warmer suitable for transfer to and from the operating theatres. These systems are integrated with mounted ventilators, monitors, infusion pumps, gas cylinders and uninterrupted battery supported power supply.

Vital signs are documented hourly, including BP (IAL), heart rate, respiratory rate, oxygen saturations, and capillary refill time. Frequency of other post-operative observations is guided by the usual management of peri-operative neonates including regular temperature assessment, pain assessment and monitoring of urinary output.

Babies with congenital heart disease are also monitored with near infra-red spectroscopy peri-operatively.

The femoral pulses should be felt initially and then again whenever assessing the baby for any concerns, particularly if there is a metabolic acidosis or rising lactate.

Differential cyanosis (pink upper extremities with cyanotic lower extremities) may occur when right-to-left shunt across a patent ductus arteriosus provides flow to the lower body. Although often not obvious to the eye, differential cyanosis may be documented based on pre ductal (right hand) and post-ductal (lower limb) pulse oximetry measurements and careful inspection. However, in the presence of lesions with large left-to-right shunt (eg, VSD), pulmonary artery saturations may approximate aortic saturations with less obvious

differential oximetry findings. For patients with Coarctation pre and post ductal oxygen saturation measurements are recommended to help assess blood flow post the lesion.

Blood pressure

Blood pressure should be monitored continuously via intra-arterial line. Where access is an issue, the blood pressure should be monitored 2-4 hourly non-invasively. Post-operatively once the arterial line is removed, non –invasive blood pressure monitoring should be measured in the lower limbs to indicate adequacy of repair at least once a shift for the first 48 hours after repair.

PGE_1

Ductal patency is maintained with a continuous infusion of PGE_1. When the neonate presents in cardiogenic shock, where the ductus is small or restrictive, a starting dose of 50 nanograms/kg/min is used. This dose may then be reduced to 10- 20 nanograms/kg/min once the ductus has opened and the clinical condition has improved. This dosing allows maintenance of ductal patency whilst minimising the generally hypotensive effect of PGE_1 and also helps to maintain tissue integrity at the preoperative site. Where there are no signs of obstruction and it is desired to keep the ductus open, a smaller starting dose of 5-10 nanograms/kg/min is selected in order to avoid apnoea in the non-ventilated patient. Doses greater than 50 nanograms/kg/min are not useful and impair tissue integrity at the surgical site.

Clinical considerations when caring for an infant on PGE_1 are:

- PGE_1 causes depression of the respiratory system and can cause apnoea.
- Apnoea is more frequent with the use of doses higher than 15nanograms /kg/min of PGE_1 [9].
- PGE_1 is given as continuous infusion and should run through a dedicated form of intravenous access (e.g. dedicated lumen of central venous access or dedicated cannula) which should not be interrupted or used for other medications or intravenous fluids. The neonate should have a second peripheral intravenous cannula to ensure the ability to provide continuity of the PGE_1 infusion if the initial cannula tissues.

Mechanical ventilation

- Invasive mechanical ventilation may be instituted to manage severe cardiac failure. Providing respiratory support takes some work off the failing heart and also allows larger dosing of the PGE_1 required to open the physiologically closed ductus arteriosus.
- Once infants are ventilated, infants' blood gases are usually attended every hour until the lactate and ventilation parameters are more stable. Preoperatively and subsequently, ABG is reviewed 4^{th} hourly to ensure stability of support.
- Non-invasive ventilation such as midline CPAP or Humidified High Flow Nasal Cannula (HHFNC) is used in patients who are more stable but may present with mild to moderate increased work of breathing. This may be due to ductal overflow into the lungs as pulmonary vascular resistance decreases.
- Ventilation parameters should be adjusted to achieve normal blood gas values.

Inotropes

Intravenous inotropes may be required in the presence of ventricular dysfunction. The inotrope and dosing regimen will be determined by the Neonatologist.

Fluid management

Postnatally diagnosed Coarctation may be associated with congestive cardiac failure and as systemic flow has been limited, hypoglycaemia is often present. Whilst it is prudent to limit intravenous fluids to around 60 mls/kg/day, this may require individual adjustment. Frusemide is also useful to manage fluid overload. Some care is required with potassium administration where there is concomitant renal impairment with elevated potassium initially and then potassium depletion associated with diuresis and correction of metabolic acidosis. Electrolytes are reviewed at the time of blood gases in the ventilated patient group.

Feeding

Caution should be exercised with enteral feeding in the pre-operative and immediate post-operative phase because of the likelihood of fluctuations in gut perfusion [10].

- Babies with an antenatal diagnosis of coarctation where the ductus is maintained from birth may be offered colostrum and comfort breast feeds.
- Babies who present with acidosis have had significant gut malperfusion and should not have enteral feeds until they are stable post-operatively because of the elevated risk of necrotising enterocolitis [10]. Mouth care with breast milk is advised. See the Immune supportive oral therapy guideline for more information.

Preparation for Theatre

The neonate should be transferred on the open care transport system to the operating theatre. It is the nurse's responsibility to check that the transport system is in good working order and perform the following checks:

- The Oxygen and Air Cylinders must be checked for adequate levels (>10, 000kpA)
- A re-breathing bag and mask is attached to the transport systems blender

Please refer to GCNC Guideline Transfer of a Neonate to Operating Theatre and other Hospital Investigative Departments (Document number 2008-0000).

Surgical Management of Coarctation

The most common surgical procedure for correction of Coarctation of aorta is resection and end to end anastomosis. This procedure consists of resecting the Coarctation and anastomosing (*connecting*) the proximal and distal aorta [12].

Post-Operative Management

In the immediate post-operative period of 12-48hours, the neonate will remain mechanically ventilated for the provision of analgesia and cardiorespiratory support. For further information on how to care for an intubated and ventilated patient refer to GCNC Practice Guideline: Respiratory Support in Neonates - GCNIC - CHW (Document No: 2007-0005).

On return from the operating theatre the following should be attended to:

Airway assessment

- Endotracheal tube size and position at the nares.
- Equal and clear air entry, on auscultation.

Blood gases

Blood gases are performed 1-4th hourly for the first twenty-four hours as guided by the neonatologist post-operatively aiming for normal values:

pH	7.35 – 7.45
PaO_2	60 – 80mmHg
$PaCO_2$	35 – 45mmHg
Base Excess	-5 – 5mmol/L
HCO_3	18 – 22mmol/L
SpO_2	92 – 94%
Lactate	<3 mol/L

Chest X-ray

To confirm the position of the endotracheal tube as well as exclude pleural fluid collections, pneumonia, and/or pneumothorax. It is also important to assess cardiac fluid status by reviewing the heart size and lung plethora.

Blood tests

- Full blood count
- Electrolytes including calcium, magnesium and phosphate
- Coagulation screen if coagulation was abnormal or required correction

Monitoring

- Systemic oxygen saturations are continuously monitored using pulse oximetry.
- Heart-rate and rhythm is monitored continuously and assessed.
- Continuous intra-arterial monitoring is continued until extubation. If the blood pressure is elevated, a nitroprusside infusion can help with afterload reduction. Non-invasive BP is checked 8 hourly to ensure correlation.
- Signs of poor systemic perfusion are oliguria, metabolic acidosis and myocardial dysfunction.

- Renal dysfunction can occur in those postnatally diagnosed secondary to pre-operative hypoperfusion & ischaemia of the kidneys. Vigilant fluid balance assessment is essential.
- The lactate level should be monitored closely. If lactate is not improving or starts rising >3mmol/L, other signs of distal hypoperfusion should be assessed such as the quality of the femoral pulses. If there is any concern the neonatologist should be informed. This may be a sign postoperatively of evolving necrotising enterocolitis.
- If low cardiac output persists, inotropic support may be required and the intravascular volume and the presence of anaemia should be reassessed.

Antibiotics

- Infants in NICU with intravenous catheters and those intubated are at increased risk of sepsis.
- Antibiotic prophylaxis is provided by intravenous cephazolin administration following surgery until removal of the chest drain and during periods of suspected sepsis.

Systemic hypertension after repair of coarctation

Post-operatively, especially in older neonates, there may be a period of hypertension secondary to an increase in release of noradrenaline during the aortic cross clamping, and for a more prolonged time from the release of renin that occurs following improvement in the perfusion of the kidneys [8].

Early post-operative control of hypertension protects the aortic anastomosis and minimises the risk of aneurysm formation in the post-stenotic segment.

Post-operative systemic arterial hypertension is managed in the first instance by adequate analgesia and sedation. If control is not readily established, then vasodilators such as sodium nitroprusside may also be used as guided by the neonatologist. Specific targets for the mean arterial pressure will be defined but generally a mean arterial pressure of 50-60 mmHg is acceptable.

Fluid and electrolytes

Intravenous fluids will be prescribed according to the blood chemistry and the fluid balance.

Chest Drain Management

A chest drain is always inserted into the left pleural space at the end of the surgical procedure. The drain assists in the drainage of collected blood, pleural fluid, and allows re-expansion of the lungs. The underwater seal drainage chamber (Atrium) should be connected to wall suction with the dry suction valve on the drainage chamber set at -20 cmH_2O unless otherwise specified by the surgeon.

Refer to SCHN Chest Drain Practice Guideline for further information and management (Document No: 2015-9017)

Pain Management

Please refer to the GCNC Pain Management in Newborns Practice Guidelines for further pain management (Document No: 2006-0028)

Dressings/Wound sites

- The dressings/wound sites are monitored and assessed each shift.
- Dressing site, drainage and integrity of the wound sites and dressings is documented in the assessment chart in the electronic medical record.
- Usually the dressing will remain intact until day five post-operatively or until discharge, whichever occurs first.
- If there are concerns with the wound or drain sites please contact the cardiothoracic fellow on-call for review.
- For preterm infants if non-absorbable sutures are utilised, ensure the date for removal is documented in the electronic medical record (usually 10-14 days).

Fluid and electrolytes

Strict fluid balance documentation is required. Normal urine output is 1-2 mL/kg/hour. If the urine output is decreased, the fluid status of the baby should be assessed by the clinical team. There may be a requirement for more or less fluid or diuretics if there is clinical evidence of fluid overload. Frusemide is the diuretic of choice and is given via IV bolus or oral dose at 1mg/kg/dose.

When given orally, frusemide is often combined with spironolactone as it has a potassium sparing effect. Spironolactone is also usually given at dose of 1mg/kg/dose. Diuretics may be given once, twice or three times a day depending on clinical requirement. Occasionally a continuous infusion of frusemide is required.

Enteral Feeds

Clinical signs of necrotising enterocolitis need to be actively assessed for due to the association with congenital heart disease and the increased risk in those diagnosed postnatally where distal perfusion (gut) has been compromised [13].

- Feeds are generally commenced in liaison between the Neonatology, cardiology and cardiac surgery teams.
- The feeds when commenced are graded up as tolerated by the infant (refer to Feeding the High-Risk Neonate - GCNC - CHW (Document No: 2007-0002)
- Neonates receiving diuretic therapy in the GCNC do not usually exceed a total fluid requirement of 150mL/kg/day. Where this is insufficient for appropriate weight gain, the feeds are supplemented with additional calories.
- The infant is weighed three times weekly as per GCNC protocol and head circumference and length are measured weekly to assess adequate weight gain in the post-operative period.
- In the instance where insufficient breast milk is available from the mother, consideration should be made for the provision of Pasteurised Donor Human Milk (PDHM) aligning with local guidelines.
- Immune Supportive Oral Therapy with mother's breast milk can be offered throughout as guided by the Neonatologist.

Complications

Vocal cord dysfunction

Following repair of aortic coarctation, there is a risk of recurrent laryngeal nerve injury and subsequent transient or permanent vocal cord palsy. Routine repair via a left thoracotomy carries a low risk (1-5%) [14] and does not require routine screening. If there is a persistently soft cry following extubation, then the infant should be referred to speech therapy and ENT for assessment including a nasendoscopy. If vocal cord dysfunction is confirmed, then a speech pathology review and modified barium swallow is indicated in order to determine the timing and method of oral feeding.

Systemic hypertension

Systemic hypertension is common but usually transient in infants who have had a coarctation repair. Persistent hypertension may require specific drug treatment guided by the cardiology team.

Other complications

- Other post-operative complications following coarctation repair are generally rare.
- Major bleeding is rare, however careful monitoring of chest drain output is essential and if there is significant bleeding (>2ml/kg) this may require further surgical intervention and blood transfusion.
- Wound infection following coarctation repair is a risk that requires close monitoring and early management. Any signs of infection such as wound erythema, swelling or purulent discharge require FBC, wound swab for culture and notification of senior staff. Due to this risk neonates in GCNC, following coarctation repair are prophylactically treated with IV cephazolin until the chest drain is removed.
- Re-coarctation is the most common long-term complication following repair of aortic coarctation. Late aneurysm formation can also occur, and for these reasons, life-long monitoring is recommended following repair.
- Chylothorax can develop postoperatively rarely due to intra-thoracic surgery

Discharge

Discharge is coordinated once feeding is established with adequate weight gain. An echocardiogram, chest X-ray and ECG are routinely performed prior to discharge.

Infants are routinely followed by the cardiac surgeon post discharge. They are referred for follow up to the Cardiac Clinical Nurse Consultant team if they are having trouble gaining weight or have other issues not specific to CoA repair. Infants may be referred to the GCNC Acute Review Clinic (ARC) for ongoing surveillance of feeding and weight gain.

The majority of neonates post coarctation repair are discharged home on no medications; occasionally diuretics (frusemide and spironolactone) are still required pending outpatient cardiology review in six weeks. Very occasionally, a baby may require beta blockers. It is the responsibility of the healthcare team to provide education for the parents regarding medications on the correct dosage and administration of these medications prior to discharge.

Following surgery in the neonatal period these infants are followed up in the Grace Centre Developmental clinic. Whilst the blood pH nadir and lactate do not correlate with neurodevelopmental delay in babies studied recently in our unit who presented postnatally they may have developmental needs that require support [15].

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