COARCTATION REPAIR IN NEONATES - GCNC

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 establishes the diagnosis and associated anatomy.
- Pre-operative management consists of providing appropriate respiratory and circulatory support, maintaining ductal patency with prostaglandin and attending to any associated fluid and electrolyte abnormality. Enteral feeding I ordinarily deferred.
- Post-operative management includes a period of mechanical ventilation, control of systemic hypertension, pain management, fluid and electrolyte balance and vigilance for immediate post-operative complications.
- There are a number of potential short term complications such as necrotising enterocolitis and vocal cord dysfunction which should be considered.

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CHANGE SUMMARY

- Document updated to include clinically relevant information including ventilation and blood pressure management
- Additional information included regarding presentation types and required treatment
- Vocal cord palsy information included
- References updated

READ ACKNOWLEDGEMENT

All clinical staff working in Grace Centre for newborn 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 can occur anywhere in the aorta, but it is most commonly found in the segment just after the 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 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 Care, suspected coarctation usually presents in 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 associated with ductal closure.

Suspected Coarctation where ductal patency is not maintained with prostaglandin

In this situation the presence of Coarctation may have been suspected, usually as a result of antenatal ultrasound, however, there are no clinical or laboratory signs of impairment of distal blood flow and intravenous prostaglandin is not used to maintain ductal patency. 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 may be undertaken 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 the index of suspicion is sufficiently low to discontinue prostaglandin and proceed according to the previous pathway.

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 around 3-10 days. The location and degree of constriction will determine the severity and timing of the presentation. Less critical Coarctation may present outside 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 renal and hepatic impairment. Since this presentation is similar to other causes of shock, including sepsis, an active

praecordium and marked hepatomegaly are valuable clinical indicators. Chest X-ray may show cardiomegaly and pulmonary plethora.

Many texts refer to upper limb hypertension and blood pressure differential between the right upper limb and the lower limbs as being an important finding [6], however, in the early acute presentation with shock where all pulses are diminished this is not a reliable differentiating sign and its absence should not exclude the possibility of left heart obstruction.

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 echocardiogram confirms the diagnosis, defines the anatomy of Coarctation and delineates any associated lesions such as ventricular septal defect and mitral valve or aortic valve anomalies.

Once the diagnosis of Coarctation requiring surgical repair is established the pre-operative management consists of regular observation of clinical signs, maintenance of ductal patency, respiratory and cardiovascular support if required and completion of those tasks required before surgery.

Initial investigations

Infants with suspected or proven Coarctation should have a chest X-ray performed along with a full blood count, electrolytes, urea and creatinine. A group and hold pending cross-match should be included as well as a CGH microarray, particular in female infants, because of the association with Turner's syndrome [7]. Coagulation studies and liver function tests should be performed if there is evidence of significant obstruction. Newborn screening should be collected pre-operatively in case blood transfusion is required.

Vital signs

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 an uninterruptable battery supported power supply.

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

The femoral pulses should be felt initially and then again whenever assessing the baby for any concerns and 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 preductal (right hand) and postductal (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 oximetric findings. For patients with Coarctation pre and post ductal oxygen saturation, measurements are recommended to help assess blood low post the lesion.

Blood pressure

Regular blood pressure observations are essential and should be taken from the right upper limb since, in the presence of aortic obstruction, blood pressure measured beyond the level of obstruction may under read.

If invasive blood pressure is required a right radial arterial line is preferable and will obviate the need for placement of a new line at the time of surgery. Non –invasive blood pressure monitoring should be undertaken in the other limbs to indicate adequacy of repair at least once a shift for the first 48 hours after repair.

Prostaglandin

Ductal patency is maintained with a continuous infusion of prostaglandin E1. Where presentation is in cardiogenic shock, where the ductus is small, a starting dose of 50 nanograms/Kg/min is often used. This dose may then be reduced to around 20 nanograms/Kg/min once the ductus has opened and the clinical condition has improved. Where there are no signs of obstruction and it is desired to keep the ductus open a smaller starting dose of 10-15 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.

Clinical considerations when caring for an infant on Prostaglandin are:

- Prostaglandin causes depression of the respiratory system and can cause apnoea.
- Apnoea is more frequent with the use of doses higher than 15 nanograms per kilogram per minute of Prostaglandin [8].
- Prostaglandin is given as continuous infusion and should run through a 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 Prostaglandin infusion if the initial cannula tissues.

Mechanical ventilation

- Mechanical ventilation may be instituted as a supportive measure for those infants presenting with severe cardiac failure. In addition to providing respiratory support ventilation overcomes the problem of apnoea associated with higher doses of prostaglandin.
- Ventilated infants should have regular, at least 4 hourly, blood gases in order to target ventilation parameters and monitor metabolic acidosis and lactate.
- 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 in consultation with cardiologist.

Fluid management

Since Coarctation may be associated with congestive cardiac failure it is usual to limit intravenous fluids to around 60 mls/Kg/day. Frusemide may also be used to manage fluid overload. Some care is required with potassium administration since there may be initial

renal impairment with elevated potassium followed by a marked fall in potassium associated with diuretic use and correction of metabolic acidosis. Frequent electrolytes should be performed at the same time as 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 alterations in gut perfusion[9].

- 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 stable post-operatively because of the elevated risk of necrotising enterocolitis [10].

Preparation for Theatre

The neonate should be transferred on the 'George' 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 transports systems blender

Please refer to GCNC Guideline Transfer of a Neonate to Operating Theatre and other Hospital Investigative Departments (No.0/C/08:0000).

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 (*remove part of*) the Coarctation and anastomosing (*connecting*) the proximal and distal aorta [11].

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 the NICU (No: 0/C/07:0005).

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

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

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

Chest X-ray

- To confirm the position of the endotracheal tube as well as excluded a large pleural fluid collection and/or pneumothorax.

Blood tests

- Full blood count
- Electrolytes
- Coagulation if coagulation was abnormal or required correction

Monitoring

- A size 8 gastric tube is inserted and aspirated initially and then placed on free drainage. The tube is aspirated every four hours. The amount of aspirate is documented in the fluid balance chart in the patient's records.
- 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. Non-invasive BP is checked 8 hourly to ensure correlation.
- Signs of poor systemic perfusion are oliguria, metabolic acidosis and myocardial dysfunction.
- Renal dysfunction is common following repair, due to pre-op ischaemia of the kidneys secondary to hypoperfusion. Vigilant fluid balance is essential.
- The lactate level should be monitored closely. If lactate is not improving or starts rising, other signs of distal hypoerfusion should be assessed such as the quality of the femoral pulses. If there is any concern the neonatologist should be informed.
- If low cardiac output persists, intravascular volume and the presence of anaemia should be reassessed. If the blood pressure permits, a low dose nitroprusside infusion can lead to afterload reduction and an improvement in metabolic acidosis.

Antibiotics

- Antibiotic prophylaxis – Intravenous cephazolin is administered following surgery until removal of the chest drain.

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 [7].

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

Postoperative 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.

Fluid and electrolytes

Intravenous fluids will prescribed according to the blood chemistry and the presence or absence of fluid overload.

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.
- Observation of the insertion site, the dressing and the amount of drainage is documented in the patient's records.
- Patency of the drain should be ensured by gently tapping and milking the drain hourly for the first 24 hours when significant bleeding can occur.
- Refer to CHW Chest Drain Practice Guideline for further information and management (No: 0/C/08:8104)

Pain Management

The Pain Assessment Tool (PAT) should be used to measure the infant's pain every two hours in the immediate post-op 24 hours. After this, the pain score is assessed every four hours for as long as the neonate is still requiring analgesics for pain relief.

Following pain assessment the appropriate actions are as follows[5]:

- Score less than 5, comfort measures should be instituted
- Score more than 10, consider an increase in analgesia
- If the score is 0-3 on two occasions and the baby is on analgesia, consider reducing the analgesia.

Please refer to the GCNC Pain Management in Newborns Practice Guidelines for further pain management (No. 1/C/06:0028)

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 including all inputs and outputs. 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.

Enteral Feeds

Clinicians should remain vigilant for clinical signs of necrotising enterocolitis because of the association with congenital heart disease [12].

- 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 GCNC *Enteral Feeding - Neonates* Practice Guideline (No. 0/C/07:0002).
- 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 weekly to assess adequate weight gain in the post-operative period.

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%) [13] 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 frug treatment according to the cardiologist.

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.

Pre-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 have any of the following:

- Low Oxygen saturations
- Post shunt insertion
- Ongoing Arrhythmias
- Pacemaker insertion
- Unrepaired cardiac defect

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 requiring 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. It is important to start this education early and reinforce the same technique each time.

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