GUIDELINE

Congenital Diaphragmatic Hernia (CDH)

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

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Read the full statement here:
CAHS Child Safe Organisation Commitment Statement

This document should be read in conjunction with this disclaimer

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Aim

To outline the presentation and management of Congenital Diaphragmatic Hernia (CDH).

Risk

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

Background

Congenital Diaphragmatic Hernia (CDH) is a developmental defect of the diaphragm that allows the abdominal contents to herniate into the chest. It occurs in approximately 1/2500 live births in Western Australia. Most defects are left sided (80-85%).

CDH is commonly classified into:

- Posterolateral (Bochdalek): most common; 70-75%.
- Anterior (Morgagni): 23-28%
- Central tendon: 2-7%.

Pathophysiology

Herniation of the abdominal contents results in ongoing hypoplasia of the ipsilateral lung and secondary mass effect leads to hypoplasia of the contralateral lung. These changes may effect:

1. **Lung mechanics**: reduced lung volumes; smooth muscle dysregulation with increased airways resistance.

2. **Pulmonary vasculature**: reduced cross-sectional area of the pulmonary bed and increased adventitial and medial increases PVR, which is “fixed” because of the chronicity and thus less responsive to vasodilators such as inhaled nitric oxide (iNO). This chronic increase in PVR may lead to right heart strain with right ventricular hypertrophy especially once the PFO and ductus closes.

Clinical Presentation

Antenatal

- Most infants with CDH will be diagnosed antenatally (80-85%)
- Right sided CDH may be missed as the liver and lung echotexture are similar
- Commonly bowel loops +/- liver are seen herniated with mediastinal shift
- Cardiac anomalies are difficult to diagnose due to the malpositioning of the heart.

Postnatal

- The classical presentation is of a newborn with severe respiratory distress, barrel chest, scaphoid abdomen with breath sounds reduced on the left and the heart sounds best heard on the right side of the chest
- About 5% of CDH will present at > 24 hours of age with mild tachypnoea, or later still with failure to thrive, recurrent chest infections, pleural effusions or as an incidental finding on CXR.

Associations

- An associated anomaly is identified in 30-50% of foetuses and 20-30% of livebirths
    - Cardiac anomalies: the most common association.
    - Genetic disorder: chromosomal rearrangements (Chrom 1, 8, 12, 15); trisomy’s’; Turner syndrome; single genes (FOG2, GATA4, FREM1); and overgrowth syndromes, Xp-linked syndromes; Donnai-Barrow; CHARGE and Cornelia de Lange.
    - Gastrointestinal disorder: duplications, atresia and malrotation.
    - Pulmonary disorder: sequestration or Congenital Pulmonary Airway Malformation (CPAM) of the Lung.

Labour and Birth Suite Management

- All antenatally diagnosed CDH infants require endotracheal intubation at birth.
- Avoid bag and mask ventilation.
- Cardiac and oxygen saturation monitoring.

- Insert a large bore nasogastric tube (8F for < 34 weeks and 10F for > 34 weeks).
- Initial ventilation goals:
  - Aim for pre-ductal SaO_2 of 85-90%.
  - Aim for the lowest PIP to achieve adequate chest wall movement and consider using rates of 40-60bpm (i.e. ventilate hypoplastic lungs using smaller, frequent breathes).

General Management and Monitoring

Each neonate will have an individualised plan that should be confirmed each day with criteria set for escalation of therapies. General cares include:

- Minimal handling; consider bolus sedation during major cares.
- Maintain normothermia, normoglycaemia, and normal electrolytes
- Monitor urine output and maintain > 1 mL/kg/hour; IDC for infants with significant sedation or muscle relaxation.
- Insert where possible a triple lumen UVC.
- Continuous invasive blood pressure and CVP monitoring.

Goals of Management

- Optimise ventilation and limit ventilator induced lung injury.
- Achieve haemodynamic stability by limiting pulmonary hypertension.
- Avoid fluid overload

Investigations

The following investigations need to be done prior to surgery:

1. Chest x-ray to confirm CDH as other conditions such as eventration or CCAM may mimic on antenatal scans. Occasionally CXR may not be able to confirm the diagnosis then consider repeat CXR, ultrasound or chest CT.
2. Echocardiography to exclude congenital heart disease and to quantify pulmonary hypertension.
3. Genetic testing: karyotype and micro-array to be done as routine. Selective testing may be considered for phenotypical syndromes such as Cornelia de Lange, Beckwith Weidermann.
4. Renal and head ultrasound to exclude any other associated anomalies.

Ventilation Support

Oxygenation and ventilation targets (without moderate-severe pulmonary hypertension):

- preductal saturation of at least 85%, but not >95%.
- arterial pCO2 between 45-60 mmHg targeting pH of 7.25-7.40.
- If a persistent leak > 25% is present with difficulty in ventilation or oxygenation, then consider upsizing the ETT.

Conventional Ventilation

- Gentle synchronised ventilation with PC-SIMV or PC-AC
- Try to maintain spontaneous respiration.
- Use volume targeted ventilation (4mL/kg); rate of 40-60bpm.
- Set maximum PIP to 25cm H_2O.
- Start with a PEEP of 5cm H_2O; if oxygenation issues and CXR shows collapse or underinflated lungs consider a trial of higher PEEP.

Consider HFOV if requiring a PIP of > 25cm H_2O to maintain pH of 7.25-7.35 and/or pCO_2 > 60mmHg. ***If an infant with CDH reaches this threshold contact the on-call fellow and/or consultant.***

High Frequency Oscillation Ventilation (HFOV)

- Commence at a MAP equal to that of CV (13-17cm H_2O).
- Consider recruitment manoeuvre then reducing MAP as tolerated to lowest level to maintain lung patency.
- Use highest frequency that will allow for adequate gas exchange.
- Delta P of amplitude adjusted until oscillation of chest is seen.
- When transitioning infant to HFOV, particularly term infants, have them well sedated and consider muscle relaxation.
- Obtain CXR within 1 hour to observe for adequate lung inflation.
    - Look at contralateral lung; 7-8 posterior ribs with a curved diaphragm.
    - If CXR shows > 9 posterior ribs or flattened diaphragm reduce the MAP.

Consider transitioning to High Frequency Jet Ventilation **(HFJV)** if:

- HFOV fails to maintain adequate gas exchange.
- HFOV is leading to cardiac dysfunction due to over distension.
- There is an air leak or gas trapping evident on CXR.

Cardiovascular support

Haemodynamic stability is influenced by multiple factors and so must be made after considering:

- Respiratory support.
- Evidence of pulmonary hypertension and right heart strain.
- Perfusion of end-organs (metabolic state, urine output and capillary refill).
- At least two standardised echocardiograms should be performed: one within 24–48 hours of life (preoperatively) and another at 2–3 weeks, to assess pulmonary hypertension and cardiac function, with additional studies as clinically indicated.

Fluid status

Pre-operative and post-operative fluid status is crucial. Monitor for signs of fluid overload (if feasible CVP persistently > 5cm H_2O; CVP trending up post fluid bolus; peripheral oedema; worsening respiratory function). If fluid overload or pulmonary congestion is present:

1. Reduce sedation and muscle relaxation to allow some spontaneous movement.
2. Judicious use of diuretics.
3. Consider fluid restriction.

Poor perfusion or Hypotension

Signs of poor perfusion:

- capillary refill >3 sec
- lactate > 3mmol/L
- urine output < 1 mL/kg/hr.

Review ventilation strategy and a trial of lower MAP may be required.

Initial management is **fluid bolus**; review after 10-20mL/kg has been given.

- May worsen RV if right heart is already under strain.
- Left ventricle is “functionally hypoplastic” and excessive fluid may precipitate pulmonary congestion.
- Serial CVP measurements and functional echocardiography are useful.

Inotropes

Consider inotropes if minimal response to fluid bolus and no evidence of excessive fluid losses. Frequent review of volume status is required prior and after commencing inotropes. CVP trends may be useful. Echocardiogram should be considered to

evaluate cardiac function and filling before and after inotropes whenever possible. There is little evidence to guide practice.

See Appendix 1: Pulmonary Hypertension Flow Chart for an overview of inotropic support in pulmonary hypertension.

a) Dobutamine
   - Consider if reduced cardiac function consider Dobutamine as first line.
   - Note: can lead to tachycardia and lead to a fall in BP however function may improve.

b) Milrinone
   - Consider if reduced function with severe PPHN
   - Note: may precipitate systemic hypotension. Hence to avoid loading and start with lower dose.
   - Has inotropy as well as may reduce pulmonary vascular resistance (PVR).

c) Adrenaline
   - Low dose adrenaline will provide inotropy without significantly increasing afterload.
   - High dose adrenaline will increase systemic vascular resistance (SVR) similar to dopamine.

d) Noradrenaline (Norepinephrine)
   - Low dose may help improve oxygenation
   - Potent pressor and will increase both SVR and PVR
   - Useful when cardiac output is normal but vasoconstriction is required

e) Dopamine
   - Low dose dopamine may have vasodilatory effect on the splanchnic vessels.
   - Medium dose dopamine will increase SVR.
   - High dose dopamine will increase both SVR and PVR.

Hydrocortisone

Consider hydrocortisone if inadequate response from fluid resuscitation and inotropic support. Send a random cortisol prior to first dose.

Inhaled Nitric Oxide

- In the presence of echocardiographic evidence of supra-systemic pulmonary hypertension without left ventricular dysfunction, inhaled nitric oxide (iNO) should be trialled if lung recruitment is adequate.
- While awaiting echocardiographic confirmation, iNO may be considered in the presence of high FiO_2 requirement with normal CO_2, together with clinical evidence of pulmonary hypertension, such as a pre- to post-ductal saturation difference >10%

- Commence iNO at 20 parts per million (ppm). Refer to Persistent pulmonary hypertension of the newborn (PPHN) and Nitric Oxide guidelines.
- Recognition of **left heart dysfunction (post-capillary pulmonary hypertension)** and the potential adverse effects of **iNO** in this physiology has increased over the past decade. iNO should therefore be used with caution when left ventricular dysfunction or elevated left atrial pressures are suspected.
- If there is no echocardiographic, clinical response, iNO should be discontinued.

Prostaglandin E1 **(Alprostadil)**

Consider PGE1 if there is severe pulmonary hypertension, particularly if there is evidence of right heart strain with a restrictive or closed ductus. By maintaining ductal patency it allows a bypass for the RV to off-load and reduces the RV afterload. This is useful especially when there is significant RV dysfunction/failure.

- Consider echocardiogram after PGE1 to document RV flow and function.
- Start at 10ng/kg/min and titrate to clinical response or echo findings.

Sildenafil

Consider in infants with refractory pulmonary hypertension where iNO has not been effective in reducing oxygen index or as an adjunct when weaning iNO and ventilatory support.

- IV or oral preparation; NOTE: IV infusion may precipitate systemic hypotension.

Milrinone

Refer to above for details.

Prostacyclin **Epoprostenol (PCC Medication Monograph)**

Prostacyclin therapy may be considered as rescue therapy before, during, or after ECMO in infants with severe refractory pulmonary hypertension.

Other therapies

Consider alkalinisation aiming for pH 7.4-7.5; increased ventilation or bicarbonate infusion.

Sedation

Appropriate sedation is required in infants with moderate-severe pulmonary hypertension or unstable with variable oxygenation.

- Commence either morphine or fentanyl
- Bolus of opioid may be required for interventions.
- Midazolam infusion may be required for muscle relaxation.
- Consider paralysis (neuromuscular blocking agent: vecuronium) in severe cases.

ECMO

A Cochrane review of ECMO in severe respiratory failure (oxygenation index > 40) in near term babies, which included CDH, significantly favoured the use of ECMO. However, while the short-term outcome of CDH babies was improved, the longer term outcomes were not improved.

ECMO Considerations

- Clinical instability on maximal medical therapy.
- Absence of pre-existing major factor (cardiac, sepsis, severe VILI, genetic).
- Impression that the acute cardiopulmonary deterioration is reversible.
- Weight criteria.
- Oxygen index.

In infants fulfilling the above criteria, a multidisciplinary team (MDT) discussion should be undertaken to determine suitability for ECMO support.

Transport of a Baby with Known or Suspected CDH

Refer to the NETS WA Clinical Guidelines.

Neonates born with CDH at KEMH should first be stabilized at KEMH; subsequent transfer to PCH should take place when the KEMH, PCH and NETS WA neonatologists consider it is safe to move the baby. Because surgery is not considered urgent it may take many days before the baby's respiratory status is stable enough to allow safe transfer to PCH.

In rare circumstances babies may not stabilise well and a meeting between the KEMH, PCH and NETS WA neonatologist and parents should occur to discuss the possible transfer of an unstable baby for surgery or ECMO. NETS WA have the capacity to transfer babies that require HFOV and nitric oxide.

Surgical Considerations

Surgery is delayed to allow for haemodynamic stability, reduction in pulmonary hypertension, appropriate ventilation and correction of any haematological or biochemical disturbances. Surgery for CDH is only available at PCH. A daily discussion with the thoracic surgeon and anaesthetist regarding the timing of surgery is required.

Surgery should be considered after 24 hours of the following:

- Adequate peripheral perfusion (U/O > 1 mL/kg/hr; lactate < 3mmol/L).
- Normal mean BP for age with no or minimal inotropic support.
- Preductal saturations > 85% -95% in FiO2 < 0.4-0.5
- Echocardiographic evidence of improved pulmonary pressures (estimated pulmonary artery pressure less than systemic pressure).

If these criteria cannot be met by 2 weeks of life, then consideration for surgery or a palliative approach should be discussed with the parents.

Babies with CDH have been operated successfully whilst still on HFOV + NO with inotropic support however a team discussion between the surgeon, PCH consultant, duty anaesthetist and parents should occur.

Ideally surgery should be delayed until after ECMO however, if unable to wean by 1 week, then surgery on ECMO or a palliative approach should be discussed with the parents.

In circumstances where the baby has not responded and continued to deteriorate, despite maximal therapies being instituted, a palliative care approach may need to be considered.

Surgery in the NICU

- Currently, CDH repairs are performed within the NICU environment (3B PCH) to allow continuity of established ventilatory and haemodynamic support without the need for transfer to theatre. **A neonatal consultant** must be present during the procedure in addition to the anaesthetic and surgical teams. See Surgical Procedures Performed on PCH NICU guideline.
- For all intraoperative CDH repairs, inhaled nitric oxide should be readily available on standby, including for infants not receiving iNO preoperatively, due to the risk of sudden increases in oxygen requirement and pulmonary hypertensive instability during surgery.
- For infants undergoing repair with ongoing high respiratory or haemodynamic support requirements, particularly those approaching or beyond 2 weeks of age, early discussion with the PICU team is recommended and PICU should be alerted prior to surgery regarding potential postoperative escalation needs.

Post-Operative Care

Post-operative handover between the NICU, anaesthetic and surgical team should be completed once the infant has returned to ward 3B. Any specific post-operative management guidelines should be raised at this handover.

All babies should have a **post-operative CXR** to confirm ET tube position, drain position, line position and expansion of the lung.

- Post-operative the ipsilateral hemithorax will be air filled.

- Over the first few days this will fill with fluid and the lung will gradually expand.
- If the infant is dependent on the ventilator after 1 week or there is cardiovascular compromise, then a small amount of fluid may need to be removed. Send this off for chylomicrons.
- Any major post-operative interventions should be discussed with the thoracic surgeon unless in an emergency situation.

Continue all supportive care as per pre-operative stabilisation management. The majority of infants will have an improvement over the first 24-48 hours post-operation and many of the supportive measures can be weaned. Extubation to nCPAP is well tolerated.

Prolonged sedation, muscle relaxation or paralysis may delay extubation and increase complications such as atelectasis or lung collapse, ventilator associated pneumonia, and poor muscle tone.

Enteral feeds can be commenced after discussion with the surgical team. Intestinal ileus and reflux is common and continuous feeds may be required in the first few days.

Discharge Planning and Follow-up

Infants who have had a CDH repair, especially those that required a patch or require supplemental oxygen at 44 weeks corrected age are at risk of multiple medical issues. The most common are poor weight gain, feeding issues and reflux. Later they may have a mixed restrictive/obstructive lung disease. About 1/3 of infants will develop scoliosis as a young adult.

The following should be arranged **prior** to discharge:

- Neonatal developmental follow-up programme (4, 8, 12, 24 months).
- Surgical follow-up (discuss timing with surgical team).
- Respiratory referral for outpatient follow-up.
- Audiology referral.
- Dietetics and feeding team as required; gastroenterology may be required for severe reflux disease.
- If post-natal echocardiogram continues to show pulmonary hypertension, then discuss follow-up with cardiologist.
- General paediatrician if a complex management plan is required.

Related CAHS internal policies, procedures and guidelines

Neonatal Medication Monographs

NETS WA Clinical Guidelines

Neonatology Guidelines

- Congenital Pulmonary Airway Malformation of the Lung
- Extra Corporeal Membrane Oxygenation (ECMO) for the Neonate
- High Frequency Jet Ventilation
- High Frequency Oscillatory Ventilation
- Post-operative Handover
- Surgical Procedures Performed on PCH NICU

References and related external legislation, policies, and guidelines

1. Wren JT Jr, Hyland RM, McNamara PJ, et al. Initial hemodynamic phenotypes and clinical trajectory in congenital diaphragmatic hernia: a pilot study. *J Pediatr.* 2026;293:115044. doi:10.1016/j.jpeds.2026.115044.
2. Puligandla P, Skarsgard E, Baird R, Guadagno E, Dimmer A, Ganescu O, et al. Diagnosis and management of congenital diaphragmatic hernia: a 2023 update from the Canadian Congenital Diaphragmatic Hernia Collaborative. *Arch Dis Child Fetal Neonatal Ed.* 2024;109(3):239-252. doi:10.1136/archdischild-2023-325865.

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

Document Owner:	Neonatology		
Reviewer / Team:	Neonatology		
Date First Issued:	June 2006	Last Reviewed:	May 2026
Amendment Dates:		Next Review Date:	May 2029
Approved by:	Neonatology Coordinating Group	Date:	26th May 2026
Endorsed by:	Neonatology Coordinating Group	Date:	
Aboriginal Impact Statement and Declaration (ISD)		Date ISD approved:	19th May 2026
Standards Applicable:	NSQHS Standards: Child Safe Principles: 1, 2, 3, 4, 5, 6, 7, 8, 9, 10		

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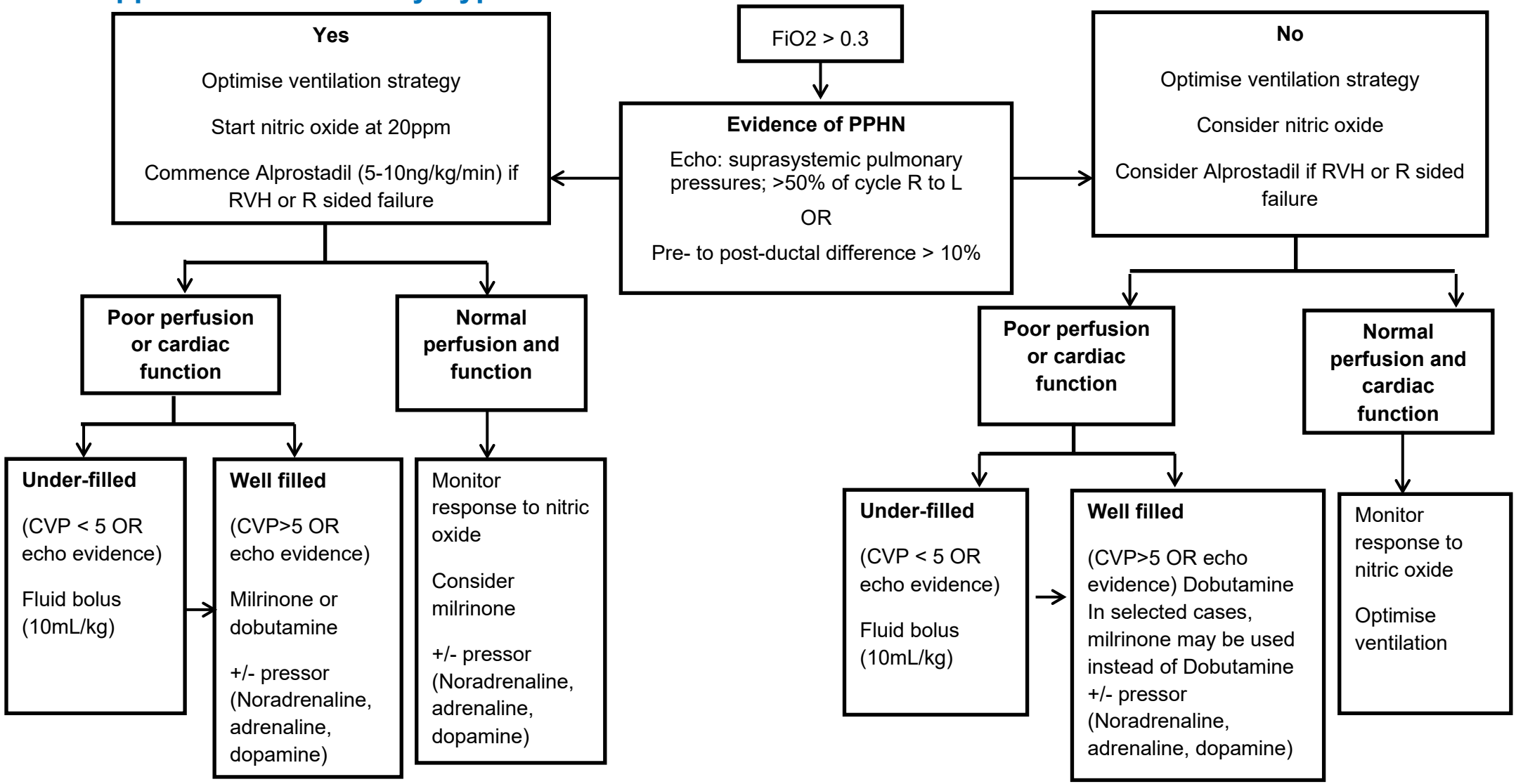
Appendix 1: Pulmonary Hypertension Flow Chart



N.B. Ensure adequate intravascular volume prior to commencing inotropes; dopamine at high doses may have greater pulmonary vasoconstriction versus peripheral vasoconstriction thus worsening pulmonary hypertension

Appendix 2: Congenital Diaphragmatic Hernia Images

(a)

Left CDH. ETT and UAC are in good position. Difficult to say where the UVC ends ?Liver. Note no gastric tube resulting in significant aeration of intra thoracic bowel loops.

(b)

Lateral of above patient.

(c)

Right CDH.

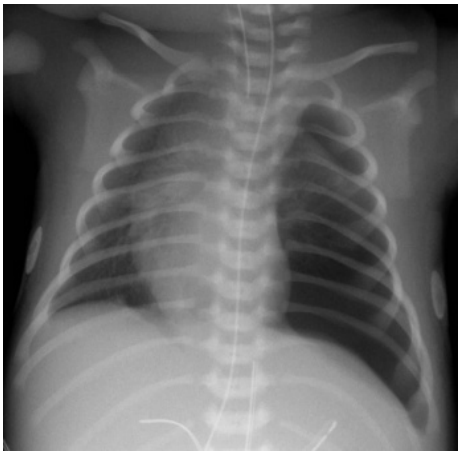
(d)

Post op CXR of above Right CDH patient. Note obvious pneumothorax.

(e)

Post op Right CDH as above. Pneumothorax filling up with fluid.

(f)



Immediate post op Left CDH. Looks like an obvious Left tension pneumothorax. Doesn't require an ICC.