GUIDELINE

Congenital Pulmonary Airway Malformation (CPAM) of the Lung

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

Aim

- Identify the etiology and epidemiology of congenital pulmonary airway malformation.
- Review the management options available for symptomatic and asymptomatic congenital pulmonary airway malformation.

Risk

CPAM has a varying clinical presentation and failure to follow the systematic approach may increase the risk of infants being managed optimally.

Definition

The new nomenclature for this condition is congenital pulmonary airway malformation (CPAM); Henceforth it will be referred as CPAM. Congenital Pulmonary Airway malformation is an abnormal airway pattern which occurs during lung branching morphogenesis. The mass of cysts lined by proliferating bronchial or cuboidal epithelium with intervening normal portions of lung.

Background

It is the most common (30-40%) congenital lung malformation. 20% may have associated congenital abnormalities. CPAM is mainly associated with other pulmonary conditions such as sequestration. CPAM is most associated with Tetralogy of Fallot.

Of CCAMs diagnosed antenatally, up to 40% may show regression in size of the cysts as the pregnancy progresses.

Pathophysiology

The hypothesis is that CPAM occurs when there is failure of interaction between endoderm and mesoderm tissue leading to an imbalance with increased cell proliferation and reduced cell death. This leads to abnormal development of the lung. New hypotheses suggest that CPAM may be part of a unifying pathogenesis that would also include sequestration, bronchial atresia and lobar emphysema. Histological studies of lung malformation suggesting both functional and organic obstructive events may lead to this condition.

Classification

At present we are still using the modified Stoker classification which is based on the histology of lung lesions:

1. Type 0 – acinar dysplasia
2. Type I – cysts up to 10cm. The cysts are lined by pseudostratified ciliated cells that are often interspersed with rows of mucous cells. Most common form.
3. Type II – Sponge-like multiple small cysts (<2cm) and solid pale tumour-like tissue. Occurs in 40% of patients. Most anomalies are associated with type II.
4. Type III – Solid. Excess of bronchiolar structures with cuboidal lining. It is localised to one area. On histology there may be evidence of hamartomatous tissue.
5. Type IV – cysts up to 10cm. The cysts are lined by flattened epithelium resting on loose mesenchymal tissue.

Clinical Presentation

CPAMs present in 3 main ways:

1. Antenatal diagnosis (symptomatic and asymptomatic)
    - Sonographic detection is maximum during second trimester (Mean GA 21-24 weeks)
    - Antenatal course may vary on ultrasound. The lesion resolves in 11-49%, regresses in 18-42% and progress in 33-44%)
    - In severe types associated complications such as mediastinal shift, hydrops and hydramnios can be detected and further imaging such as MRI may be warranted.
2. Symptomatic post-natal diagnosis
3. Asymptomatic post-natal diagnosis (incidental on CXR).

- Common symptoms and signs are early tachypnoea and poor feeding. Most infants are asymptomatic and present later with tachypnoea, repeated chest infections, bronchiectasis and rarely lung abscess.
- Approximately 20% have associated malformations of which the majority are other pulmonary associations. Other associations include renal and cardiac.
- Long term prognosis depends on whether the infant becomes symptomatic with recurrent infections prior to surgical removal. Emergency surgery carries the highest risk. The majority that are asymptomatic at birth will remain asymptomatic until surgery.

Investigations

Often made antenatally, with the differential being CDH or sequestrated lung. Pulmonary sequestration, is identified by the visualization of a vessel coming directly from the aorta in Doppler

- Postnatally the CXR should be done on day 1 of life
    - If the infant is asymptomatic delay the CXR until the infant is over 12 hours old. This allows for fluid resorption to occur and improves interpretation of the CXR.

Management

For antenatal diagnosis, refer to antenatal management plan for early resuscitation and post-natal management.

Symptomatic

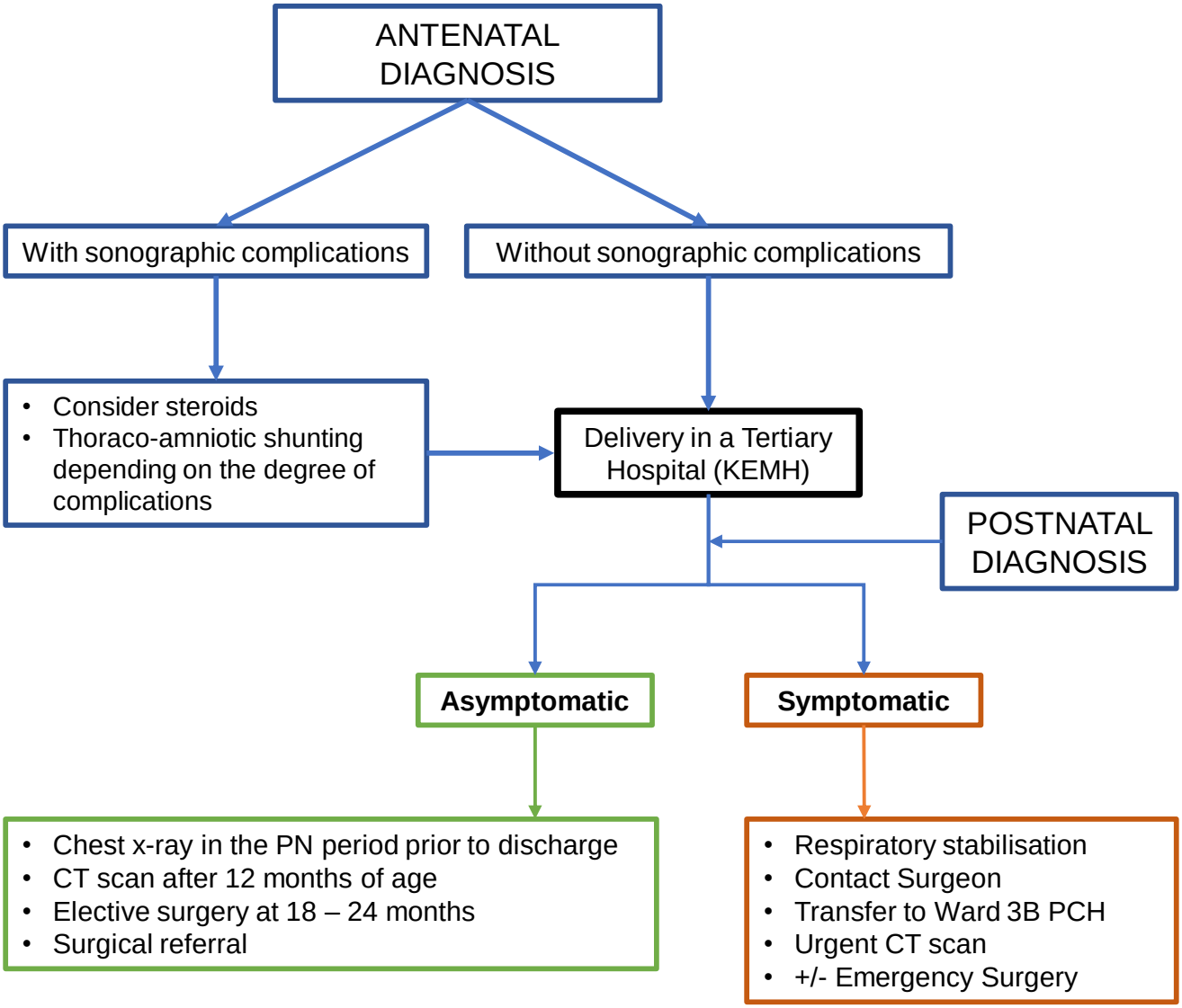
If the infant is symptomatic at birth, then admit to the neonatal unit and perform an early CXR. Discuss acute management with the neonatal consultant and the surgical team at PCH.

If the symptoms are related to the CPAM rather than normal newborn respiratory disease, then the baby should be transferred to PCH for evaluation. Infants will likely require a CT chest +/- lobectomy if the baby remains symptomatic.

Asymptomatic

If the infant is asymptomatic then the baby can be managed on the postnatal ward. Perform a CXR on day 1-2 of life and contact the surgical team for management. Most infants can be discharged home with outpatient follow-up and CT scan.

Most infants have the CCAM excised by 3 months -2 years of age because of the increased risk of adenoma and increased risk of infections.


ANTENATAL DIAGNOSIS
With sonographic complications
Without sonographic complications
• Consider steroids
• Thoraco-amniotic shunting depending on the degree of complications
Delivery in a Tertiary Hospital (KEMH)
POSTNATAL DIAGNOSIS
Asymptomatic
Symptomatic
• Chest x-ray in the PN period prior to discharge
• CT scan after 12 months of age
• Elective surgery at 18 – 24 months
• Surgical referral
• Respiratory stabilisation
• Contact Surgeon
• Transfer to Ward 3B PCH
• Urgent CT scan
• +/- Emergency Surgery

References

1. Leblanc C, Baron M, Desselas E, Phan MH, Rybak A, Thouvenin G, Lauby C, Irtan S. Congenital pulmonary airway malformations: state-of-the-art review for pediatrician's use. Eur J Pediatr. 2017 Dec;176(12):1559-1571.
2. Wong KKY, Flake AW, Tibboel D, Rottier RJ, Tam PKH. Congenital pulmonary airway malformation: advances and controversies. Lancet Child Adolesc Health. 2018 Apr;2(4):290-297.

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Document Owner:	Neonatology		
Reviewer / Team:	Neonatology Coordinating Group/Surgical		
Date First Issued:	June 2006	Last Reviewed:	May 2022
Amendment Dates:		Next Review Date:	24th May 2025
Approved by:	Neonatology Coordinating Group	Date:	24th May 2022
Endorsed by:	Neonatology Coordinating Group	Date:	
Standards Applicable:	NSQHS Standards:  Child Safe Standards: 1,10		

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