GUIDELINE

Congenital Pulmonary Airway Malformation (CPAM) of the Lung

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

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Aim

- Identify the aetiology and epidemiology of congenital pulmonary airway malformation (CPAM).
- Review the management options available for symptomatic and asymptomatic CPAM.

Risk

CPAM has a varying clinical presentation and failure to follow a systematic approach may increase the risk of infants receiving suboptimal management.

Definition

CPAM 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 mostly associated with Tetralogy of Fallot. Of CPAM's 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:

- Type 0 – acinar dysplasia
- 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.
- 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.
- 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.
- Type IV – cysts up to 10cm. The cysts are lined by flattened epithelium resting on loose mesenchymal tissue.

Clinical Presentation

CPAM's 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 Neonatal Management Plan MR409.90 for early resuscitation and post-natal management.

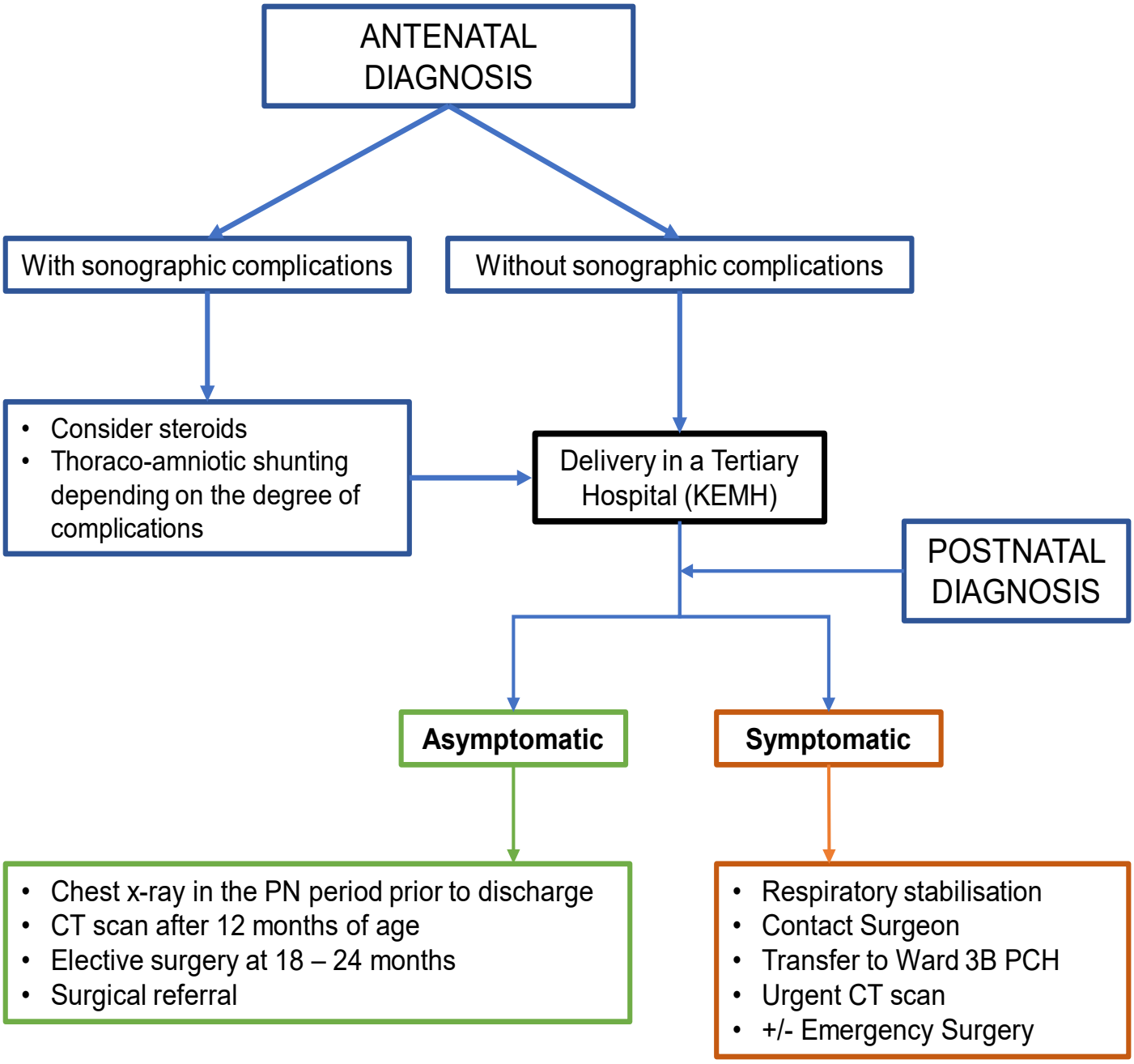
Symptomatic

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 infant should be transferred to PCH for evaluation. Infants will likely require a CT chest +/- lobectomy if the infant remains symptomatic.

Asymptomatic

If the infant is asymptomatic then the infant 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 at 3 months of age and CT scan chest with IV contrast under GA after the child is more than 12 months of age.



Justification for Surgical Management of Asymptomatic CPAM

Most infants have the CPAM excised by 3 months -2 years of age

Despite the absence of symptoms, elective surgical resection of CPAM is advocated due to the potential risk of malignancy (increased risk of adenoma) and future complications due to infections, and because early planned surgery allows for compensatory lung growth and is associated with fewer postoperative complications compared with delayed or emergency intervention.

References

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3. Cook J, Chitty LS, De Coppi P, Ashworth M, Wallis C. The natural history of prenatally diagnosed congenital cystic lung lesions: long-term follow-up of 119 cases. Archives of disease in childhood. 2017 Sep 1;102(9):798-803.

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