Management of infants with antenatally suspected congenital cystic lung lesions

‘Cystic lung lesions’ is the umbrella term for Congenital Pulmonary Airway Malformation and Bronchopulmonary Sequestration. These can be difficult to distinguish both antenatally and postnatally and some cases are “hybrid lesions”. All infants with suspected lesions will be delivered at UHW.

Congenital Pulmonary Airway Malformation (CPAM)
CPAM is a rare developmental anomaly of the lower respiratory tract, consisting of a multicystic mass of pulmonary tissue in communication with the pulmonary tree. Formation is not related to maternal factors. There is no confirmed gender bias. Incidence is in the range of 1 per 8300 to 35,000 live births [1].

<table>
<thead>
<tr>
<th>Type</th>
<th>% of CPAMs</th>
<th>Histology</th>
<th>Clinical Course</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>1-3%</td>
<td>Small cysts (&gt;5mm) involving entire lung</td>
<td>Severe impairment of gaseous exchange. Death following delivery.</td>
</tr>
<tr>
<td>1</td>
<td>60-70%</td>
<td>Single or multi-loculated cysts. Only one lung involved in 95% of cases. Normal alveoli present.</td>
<td>Clinical presentation dependent upon size of cysts (and subsequent mass effect) with larger cysts presenting earlier with more severe respiratory distress. Has malignant potential in later life.</td>
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<td>2</td>
<td>15-20%</td>
<td>Multiple cysts and solid areas blending to adjacent normal tissue.</td>
<td>Little mass effect. Can be difficult to distinguish from extralobar pulmonary sequestration. Associated congenital anomalies in 60%.</td>
</tr>
<tr>
<td>3</td>
<td>5-10%</td>
<td>Large lesions consisting of small cysts - involving entire or several lobes</td>
<td>Early neonatal presentation with severe respiratory distress. Associated with poor outcomes.</td>
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<tr>
<td>4</td>
<td>10-15%</td>
<td>Large, often single cysts</td>
<td>Tend to present later in childhood with recurrent infection or tension pneumothorax. Strong association with malignancy.</td>
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Presentation
Antenatal
- Usually diagnosed antenatally on fetal anomaly scanning: echogenic areas within the lung.
- Literature reports fetal hydrops develops in 40% of cases [3], secondary to mass effect causing mediastinal shift, and obstruction of the inferior vena cava with increased central venous pressure; pulmonary hypoplasia is likely.
- Lesions regress during the course of gestation in around 60% cases [4]. CPAMs usually demonstrate peak growth between 25-28 weeks gestation. Serial USS monitoring is required.
- **If lesion remains visible during third trimester ultrasound scanning, delivery at tertiary unit is advisable.**
- Further details on patients will be recorded in the fetal medicine folder.

Postnatal
- **Two thirds of postnatally diagnosed CPAMs** present in the newborn period with evidence of respiratory distress. Likelihood of respiratory distress and its severity increases with lesion size.
- Signs of respiratory distress include tachypnea, increased respiratory effort – grunting, recessions, head-bobbing and cyanosis or low oxygen saturations.
- Can present as spontaneous pneumothorax in the neonatal period.
- One third present after the neonatal period, usually with recurrent pneumonia.

Management
At Delivery
- Please inform the neonatal team on call of imminent delivery. Neonatal Registrar to be present at delivery to provide respiratory support if required.
- Management as per Neonatal Life Support algorithm.

The majority of antenatally diagnosed patients with CPAM are asymptomatic at birth [5], requiring normal postnatal care. Antenatally diagnosed large CPAM size, polyhydramnios and hydrops are more likely to require immediate respiratory support.

Following delivery - inform the paediatric surgical registrar on call of delivery.

Asymptomatic infants
- Observe infants for evidence of respiratory distress: tachypnea, increased respiratory effort – grunting, recessions, head-bobbing and cyanosis. Where admission is not required, monitor infants 4 hourly; observations (heart rate, saturations, respiratory rate, temperature) for 24 hours.
- In asymptomatic infants, perform oxygen saturations after 10 minutes of age to look for evidence of impaired oxygenation.
- Normal feeding can be commenced.
- Perform a plain chest radiograph after 4 hours, on the first day of life. Can appear as single or multiple lesions, which are entirely air filled, or have air fluid levels. Can be ‘bubbly’ or homogenous. Look for evidence of hypoplasia in the ipsilateral lung (due to mass effect) and mediastinal shift. **Normal radiograph does not exclude CPAM [5].**
- Asymptomatic infants with an abnormal chest radiograph require inpatient paediatric respiratory review prior to discharge. If chest radiograph is normal, outpatient review may be appropriate. Refer via the paediatric respiratory registrar on call in both cases.
- CT of the chest with contrast at 3–4 months of age to confirm the diagnosis and further evaluate the lesion. Discuss with radiology and surgical team prior to discharge.
- Surgical resection is the definitive treatment if the lesion is significant. The timing is controversial: two retrospective cohort studies and a subsequent review have suggested that 3–6 months is the optimum age for surgery \[6, 7, 8\]. However, surgical resection later in childhood is associated with improved weight and height outcomes compared to intervention in infancy \[9\].
- Treatment of asymptomatic lesions remains controversial. If deferred, surveillance due to increased risk of pulmonary malignancy will be required with serial CT scanning \[10\] – decided on case-by-case basis by respiratory team.

On discharge, consider arranging open access via Children’s Assessment Unit. Appropriate counseling of parents regarding ‘red flag’ symptoms: respiratory distress, poor feeding, and failure to thrive.

**Symptomatic infants**

Admission to the neonatal intensive care unit for optimisation of ventilation.

Inform the neonatal consultant on call, and the paediatric surgical consultant on call of delivery.

Inform the paediatric respiratory team of the admission to the neonatal intensive care unit.

- IV access: FBC, U and E, CRP, LFT, group and save, blood gas.
- Nasogastric tube insertion – can be associated with oesophageal atresia and tracheo-oesophageal fistula.
- Commence IV fluids as per protocol and to remain nil by mouth if significant respiratory distress.
- Commence benzyl penicillin and gentamicin at weight and gestation appropriate dosing.
- Perform plain chest radiograph as soon as possible. Consider the need for early CT scanning to guide further management. MRI scanning may also be utilised in the future.

A thorough, early ‘baby check’ should be performed.

Type II CPAMs are associated with other congenital anomalies in 60% of cases, including oesophageal atresia, tracheo-oesophageal fistula, bilateral renal agenesis, intestinal atresia, diaphragmatic, cardiac and bony anomalies.

Consider array CGH if other abnormalities present; although CPAMs are not typically associated with chromosome anomalies.

Surgical resection is the definitive management. Infants should be discussed at the Thursday surgical-neonatal meeting.

**Bronchopulmonary Sequestration (BPS)**

BPS is a rare congenital malformation of the lower respiratory tract; consisting of a nonfunctioning mass of lung tissue that lacks normal communication with the tracheobronchial tree. BPS receives its arterial blood supply from the systemic circulation.

**Diagnosis**

<table>
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<th>% of BPS</th>
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<tr>
<td>Intralobar Pulmonary Sequestration (ILS)</td>
<td>75-90%</td>
<td>Lesion within the normal lobe, lacks own visceral pleura</td>
<td>Males = Females. Around 20% have associated congenital anomalies : cardiac, congenital diaphragmatic hernia, vertebral anomalies, colonic duplications</td>
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<tr>
<td>Extralobar Pulmonary Sequestration (ELS)</td>
<td>10-25%</td>
<td>Mass located outside normal lung (can be sub-diaphragmatic or retroperitoneal), own visceral pleura</td>
<td>Most commonly presents in fetal / neonatal period Male preponderance Around 40% have associated anomalies as above Usually left sided, lower lobe</td>
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Usually antenatally diagnosed. See fetal medicine folder for further information. Can regress. Expect more severe presentation with respiratory distress where there has been antenatal hydrops. Can present postnatally with respiratory distress in the neonatal period; or recurrent infection (and more rarely high output failure) later in childhood.

**Management**

as per CPAM but including

Chest radiograph may show a uniformly dense mass within the thoracic cavity / pulmonary parenchyma. Usually in lower lobes, but can be anywhere within thorax. Lesions may not be apparent on chest radiograph.

Early involvement of cardiology team – ultrasonography with Doppler imaging.

CT scanning at 3 months with contrast to define the lesion and aberrant artery. If evidence of significant Surgical excision is curative and associated with minimal morbidity. It can be performed early or later in childhood dependent upon symptoms. Occasionally serial monitoring is appropriate, with CT/MRI studies performed every 5 years in childhood.

**References**


Kate Burke, Sybil Barr and Jenny Calvert September 2014 to be September May 2017.
Pathway of antenatally detected congenital lung mass

Antenatal scans suggest congenital lung mass

CXR at least 4 hours after birth

CXR normal?

Yes

No

Respiratory symptoms?

Yes

Admit to either NICU or postnatal ward. Inform surgical and respiratory services.

No

Monitor for at least 24 hours. Respiratory service review prior to discharge. Respiratory follow up.

Monitor for 24 hours. Refer to respiratory service for outpatient review.