King's College Hospital
NHS Foundation Trust

Congenital lung malformation (CLM)

Information for parents

This information sheet answers some of the questions you may have about congenital lung malformation (CLM). It explains how CLM can be diagnosed, potential treatment and management options, and provides further reading information. If you have any other questions or concerns, please do not hesitate to speak to the doctors or nurses caring for your child.

What is CLM?

Congenital lung malformation (CLM) is a condition present at birth where part of a baby's lung develops abnormally. This can result in cystic or solid lesions that may affect normal lung function.

How common is CLM?

CLM is estimated to occur in approximately 1 in 2,500 live births. The detection rate is increasing due to improved prenatal imaging.

What are the causes of CLM?

The exact cause of CLM is unknown, but it is believed to occur due to unexpected lung tissue development during pregnancy. CLM is not hereditary (it does not run in families).

How is CLM diagnosed?

Usually, the diagnosis of a CLM can be made in one of the following 3 scenarios:

1. Antenatal diagnosis

CLM is often detected during a routine antenatal ultrasound scan. When CLM is identified during pregnancy, close monitoring is essential to track the lesion's growth and assess its potential impact on the baby's health. Frequent ultrasounds help determine whether the lesion is decreasing, is stable or is increasing in size.

In cases where the lesion is large or causing complications such as mediastinal shift (pushing the baby's heart) or fetal hydrops (abnormal fluid collections in the fetus, such as around the heart or lungs, in the abdomen, or in the skin and soft tissues), medical interventions may be considered. These may include maternal steroid therapy or, in rare instances, fetal intervention to drain or block the blood supply to the lesion. A multidisciplinary team will develop a birth plan to ensure a safe delivery and appropriate neonatal care.

2. Incidental finding on imaging performed for another reason

3. In older children, diagnosis may be made due to infections that keep coming back in the affected area of the lung

4. Adult patients may present with infections or (a lot less commonly) evidence of cancer in the affected part of the lung

What conditions can be part of CLM?

Congenital lung abnormalities can include:

- **congenital pulmonary airway malformation (CPAM) previously known as congenital adenomatoid malformation (CCAM):** abnormal lung tissue forming cystic or solid lesions
- **bronchopulmonary sequestration (BPS):** a non-functioning mass of lung tissue with its own blood supply – this can be intralobar (inside the normal lung) or extralobar (outside normal lung)
- **congenital lobar overinflation (CLO):** overinflation of a lung lobe causing breathing difficulties
- **congenital bronchial atresia:** part of the bronchial tree is blocked, leading to mucus accumulation and air trapping with overinflation of the affected lung segment
- **bronchogenic cyst:** a cystic lesion that occurs when a small portion of the developing trachea (windpipe) and lung pinch off and become separate from the airway

What are the effects on the baby at birth?

The impact of CLM varies from asymptomatic to severe. In some cases, CLM remains small and does not cause symptoms. However, larger lesions may press on nearby organs, leading to breathing difficulties at birth or soon after birth.

In rare cases a cyst can pop, releasing air outside the lungs. In this case, a tube is placed in the baby's chest to release the air.

What CLM investigations take place after birth?

After birth, babies diagnosed with CLM undergo further investigations to assess the size, location, and impact of the lesion. These may include:

- **chest X-ray:** a simple imaging test that might show lung lesions, especially if they are of a large size that move the heart to one side
- **contrast CT (computed tomography) scan:** provides a more detailed view of the lung structure, identifying smaller lesions and helps in surgical planning if an operation is needed
- **MRI (magnetic resonance imaging):** occasionally used for further assessment, especially for complex cases.
- **ultrasound:** occasionally used for further assessment, especially for complex cases.
- **echocardiography:** performed on selected cases to evaluate heart function and detect any associated congenital heart abnormalities

Treatment and management

- **Before birth:** Regular monitoring by ultrasound is essential. In severe cases, interventions such as steroid treatment or fetal intervention may be considered.
- **After birth:** Many babies with CLM show no symptoms and need only a short period of observation in the hospital. Following discharge, a contrast CT scan is organised in the first few months of life. This is followed by a multidisciplinary clinic visit where the baby is assessed, the images are discussed and a decision is reached if surgery is needed. However, if the lesion causes breathing difficulties, oxygen support or surgery may be needed soon after birth.
- **Surgical removal:** A lobectomy (removal of the affected lung lobe) or excision of the lesion (bronchopulmonary sequestration) may be recommended to prevent infections, and potential

malignancy (cancer) later in life. This will also allow further expansion and compensatory growth of the compressed normal lung.

Risks of conservative management and surgery

If a child shows no symptoms (asymptomatic), a careful assessment of the benefits and risks of surgery versus conservative management is done. This is based on the CT findings and considers parental preference. There is no risk-free option.

- **Conservative management:** Choosing to monitor CLM without immediate surgery avoids the risks of a major surgery but carries some other risks. Although some lesions remain asymptomatic, others may grow and lead to respiratory issues, infections, or even malignancy (very rare) in later life. Regular follow-ups are crucial to detect any changes early.
- **Surgical treatment:** Surgery is generally safe but carries typical risks associated with operations, such as bleeding (very rarely can be catastrophic and pose risk of death), infection, injury to nearby structures, prolonged air leak and reactions to anaesthesia. The surgery is usually performed keyhole. There may be a need to convert to open surgery in challenging cases. Most children recover well, with the remaining lung compensating for the removed tissue.

Long term follow-up

- **Conservative management**: The child will be seen in clinic every year and a repeated CT scan will be performed from the age of 5 years. Surgery will be offered if any complications arise, such as infection or if the lesion has significantly increased in size. The child will continue to be followed up in the respiratory medicine clinic.
- **Surgical treatment**: After recovery from surgery, the child will continue to be followed up in the respiratory medicine clinic.

What is the long-term outcome?

Most children with CLM lead healthy lives, especially if the condition is monitored and managed appropriately. After surgery, lung function typically improves as the remaining lung grows and compensates for the removed tissue.

Early detection, monitoring, and timely intervention can ensure the best outcomes for your baby.

Further reading

Asthma + Lung UK: Congenital pulmonary airway malformation (CPAM): www.asthmaandlung.org.uk/conditions/congenital-pulmonary-airway-malformation-cpam

MyChart

Our MyChart app and website lets you securely access parts of your health record with us, giving you more control over your care. To sign up or for help, call us on **020 3299 4618** or email **kings.mychart@nhs.net**. Visit **www.kch.nhs.uk/mychart** to find out more.

Sharing your information

King’s College Hospital NHS Foundation Trust has partnered with Guy's and St Thomas' NHS Foundation Trust through the King’s Health Partners Academic Health Sciences Centre. We are working together to give our patients the best possible care, so you might find we invite you for appointments at Guy's or St Thomas' hospitals. King’s College Hospital and Guy’s and St Thomas’ NHS Foundation Trusts share an electronic patient record system, which means information about

your health record can be accessed safely and securely by health and care staff at both Trusts. For more information visit **www.kch.nhs.uk**.

Care provided by students

We provide clinical training where our students get practical experience by treating patients. Please tell your doctor or nurse if you do not want students to be involved in your care. Your treatment will not be affected by your decision.

PALS

The Patient Advice and Liaison Service (PALS) is a service that offers support, information and assistance to patients, relatives and visitors. They can also provide help and advice if you have a concern or complaint that staff have not been able to resolve for you. They can also pass on praise or thanks to our teams. The PALS office is located on the ground floor of the Hambleden Wing, near the main entrance on Bessemer Road - staff will be happy to direct you.

PALS at King's College Hospital, Denmark Hill, London SE5 9RS
Tel: **020 3299 4618**
Email: **kings.pals@nhs.net**

If you would like the information in this leaflet in a different language or format, please contact our Interpreting and Accessible Communication Support on 020 3299 4618 or email kings.access@nhs.net

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