CONDITION

Cystic lung disease

Cystic lung disease is a condition in which abnormal air-filled spaces develop within the lungs.

WHAT IT IS
Multiple abnormal air-filled spaces within the lungs

BODY SYSTEM
Lungs and breathing

MAIN SYMPTOMS
Breathlessness, cough, chest pain, collapsed lung

WHAT CAUSES IT
Inherited, immune, smoking-related, infectious, or unknown

HOW IT IS DIAGNOSED
High-resolution chest CT and cause-specific tests

MAIN TREATMENT
Cause-directed care and management of lung complications

CAN IT BE CURED?
Sometimes - the underlying cause may be treatable

IS IT AN EMERGENCY?
Sometimes - sudden breathlessness or one-sided chest pain

LONG-TERM OUTLOOK
Ranges from stable to progressive based on the cause

Overview

Cystic lung disease is a condition in which abnormal air-filled spaces, called cysts, develop within the lungs. A lung cyst has a defined wall and is usually identified on a CT scan.

The term describes an imaging pattern rather than one specific disease. It can result from inherited, immune-related, smoking-related, infectious or other conditions. Finding the cause matters because treatment and monitoring vary.

Cysts are different from normal lung air sacs and from cavities, which are often thicker-walled spaces caused by infection, inflammation or cancer. Emphysema can resemble cysts on a scan, but its air-filled areas usually lack clear walls.

Some people have a few cysts without symptoms. Others have widespread disease that

affects breathing or increases the risk of a pneumothorax, commonly called a collapsed lung. This occurs when air leaks between the lung and chest wall.

Cystic lung disease is not the same as cystic fibrosis, an inherited disorder that causes thick mucus and affects several organs.

Symptoms

Symptoms depend on the number, size and location of the cysts and their underlying cause. Cysts may be found by chance during imaging for another reason.

Possible symptoms include:

A pneumothorax can be the first sign. Breathlessness and pain often begin suddenly, although a small pneumothorax may cause milder symptoms.

Some causes also affect other organs. Birt-Hogg-Dubé syndrome can cause distinctive skin bumps and increase the risk of certain kidney tumours. Lymphangioleiomyomatosis, or LAM, may be associated with kidney growths called angiomyolipomas and fluid around the lungs.

- Shortness of breath, especially during activity
- A persistent dry or mucus-producing cough
- Reduced exercise tolerance or unusual tiredness
- Sudden, usually one-sided chest pain from a pneumothorax
- Repeated chest infections
- Coughing up blood, which is less common

Causes and risk factors

The pattern and location of cysts on a CT scan can provide clues to the cause.

LAM mainly affects women. It results from abnormal smooth muscle-like cells growing in the lungs and lymphatic system. It may occur alone or with the inherited condition tuberous sclerosis complex.

Birt-Hogg-Dubé syndrome is caused by a change in the FLCN gene and can run in families. Lung cysts are often near the lower and inner lungs, and repeated pneumothoraces may occur.

Pulmonary Langerhans cell histiocytosis is strongly associated with cigarette smoking and involves an abnormal build-up of immune cells in the lungs. Stopping smoking is central to treatment.

Lymphoid interstitial pneumonia involves immune cells accumulating in lung tissue. It may occur with autoimmune conditions such as Sjögren disease, infections or immune system disorders.

Other causes include previous infections, light-chain deposition disease, amyloidosis and rare cancers. Emphysema and age-related changes can sometimes look similar.

Testing does not always identify a cause. A few isolated cysts may not represent progressive disease, particularly when lung function is normal.

Diagnosis

A high-resolution chest CT is the main test. It shows the cysts' walls, shapes, sizes and distribution.

Your medical history can help identify the cause, including smoking, previous pneumothoraces, autoimmune symptoms and relatives with lung collapse, kidney cancer or similar scan findings.

Further testing may include pulmonary function tests, which measure lung volume, airflow and how well oxygen passes into the blood.

Blood tests may check for autoimmune disease, infection or proteins linked to specific conditions. A test for vascular endothelial growth factor D, or VEGF-D, can support a diagnosis of LAM in the appropriate setting.

Genetic testing may be offered if an inherited condition such as Birt-Hogg-Dubé syndrome is suspected. Because results may affect relatives, genetic counselling can help explain the implications and limitations.

A lung biopsy removes tissue for examination. It is not needed for everyone but may be considered when less invasive tests do not establish the cause and the result would change care.

Treatment and management

Treatment depends on the cause, lung function and complications. Existing cysts often remain even when the underlying condition is controlled.

For LAM, sirolimus may be used in selected people with abnormal or declining lung function or certain complications. It can slow disease activity but does not remove established cysts.

For pulmonary Langerhans cell histiocytosis, stopping smoking and avoiding tobacco smoke are essential. Persistent or extensive disease may need specialist treatment.

Immune-related conditions may be treated by managing the associated autoimmune or blood disorder. The possible benefits of medicines that suppress immune activity must be balanced against their risks.

Supportive care may include pulmonary rehabilitation, oxygen for persistently low blood oxygen and vaccination against respiratory infections.

A pneumothorax may require observation, removal of air through a chest tube or a procedure to reduce recurrence. Pleurodesis joins the lung surface to the chest wall. Because pneumothoraces can recur, specialist follow-up is important.

Lung transplantation may be considered for selected people with severe, advanced disease despite other treatment.

Complications

A pneumothorax is one of the most important complications. It can recur and affect either lung.

Other possible complications include:

Complications outside the lungs can include kidney tumours in Birt-Hogg-Dubé syndrome and kidney angiomyolipomas in LAM. Screening is based on the confirmed diagnosis rather than lung cysts alone.

- Gradual loss of lung function
- Low blood oxygen
- Repeated respiratory infections
- Bleeding into the airways
- Fluid around the lungs
- Pulmonary hypertension, or high pressure in lung blood vessels
- Respiratory failure in advanced disease

Prevention

Cystic lung disease cannot always be prevented. Inherited conditions such as Birt-Hogg-Dubé syndrome and tuberous sclerosis complex are caused by gene changes.

Not smoking can prevent much of the risk associated with pulmonary Langerhans cell histiocytosis. If you smoke, stopping also protects healthy lung tissue and may improve

treatment safety.

Treating related infections or immune disorders may limit further injury, although established cysts may remain.

Pressure changes matter because cysts can increase pneumothorax risk. Do not fly with a current or untreated pneumothorax. Scuba diving may be unsafe even if you feel well. Seek specialist advice before diving or after a recent lung collapse.

Living with it and what to expect

The outlook depends on the cause. Some people have stable cysts and normal activity for years. Others experience repeated pneumothoraces or gradually declining lung function.

Follow-up commonly includes symptom reviews and breathing tests, with imaging when needed. CT scans are not necessarily repeated at every visit; timing depends on the diagnosis, previous findings and changes in symptoms or lung function.

Regular activity is usually helpful within your breathing limits. Pulmonary rehabilitation may improve fitness and confidence. A sudden loss of exercise ability should be assessed rather than assumed to be part of the condition.

Pregnancy can affect some conditions, particularly LAM, and may change the risk of pneumothorax or other complications. Before pregnancy, review your diagnosis, medicines and individual risks with your specialists.

If the condition is inherited, genetic counselling may help relatives decide whether testing is appropriate. Their risk cannot be determined from your CT scan alone.

Many people continue to work, travel and exercise with appropriate follow-up. Practical priorities include avoiding smoke, recognising pneumothorax symptoms and managing the underlying condition.

When to get medical help

Seek emergency medical care for:

These symptoms may indicate a pneumothorax, significant bleeding or another serious breathing problem. Do not drive yourself if you are severely short of breath or faint.

Arrange routine medical follow-up for a persistent cough, gradually worsening breathlessness, reduced exercise tolerance or repeated chest infections. Follow-up is also important after any pneumothorax, even if symptoms have settled.

Mention any family history of repeated collapsed lungs, kidney cancer or Birt-Hogg-Dubé syndrome because it may change which tests are appropriate.

- Sudden or severe shortness of breath
- Sudden one-sided chest or shoulder pain
- Blue or grey lips, confusion, fainting or severe difficulty speaking
- Coughing up more than small streaks of blood
- Rapid worsening after a recent pneumothorax or chest procedure

Common questions

Is cystic lung disease cancer?

Usually not. It describes an imaging pattern, not cancer itself. However, a few rare cancers or blood-protein disorders can cause lung cysts, so careful assessment is needed.

Do lung cysts go away?

Established cysts often remain. Treating the cause may slow new cyst formation, protect lung function or reduce complications. Some infection-related spaces may improve.

Is cystic lung disease the same as cystic fibrosis?

No. Cystic fibrosis is an inherited disorder involving thick mucus, repeated infections and several organs. Cystic lung disease refers to air-filled spaces in lung tissue and has many possible causes.

Can I exercise with lung cysts?

Many people can exercise safely, but suitable intensity depends on symptoms, lung function and previous pneumothoraces. Stop and seek urgent assessment if exercise causes sudden chest pain or marked breathlessness.

Can I fly with cystic lung disease?

Stable disease does not automatically prevent flying. However, cabin pressure can expand trapped air. Do not fly with an active or untreated pneumothorax, and seek individual advice after a recent episode or procedure.

Can cystic lung disease run in families?

Some forms can. Birt-Hogg-Dubé syndrome and tuberous sclerosis complex are inherited, while many other causes are not. Genetic testing is usually considered when the history or scan pattern suggests it.

Sources

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