CONDITION

Pulmonary alveolar proteinosis

Pulmonary alveolar proteinosis is a rare condition in which protein-rich material builds up inside the lungs' air sacs.

WHAT IT IS
Protein-rich material collecting inside the lungs' air sacs

BODY SYSTEM
Lungs and breathing

MAIN SYMPTOMS
Shortness of breath, cough, fatigue, reduced exercise tolerance

WHAT CAUSES IT
Usually autoimmune; sometimes secondary or inherited

HOW IT IS DIAGNOSED
Lung imaging, antibody testing, and bronchoscopy fluid analysis

MAIN TREATMENT
Lung washing, inhaled immune therapy, and supportive care

CAN IT BE CURED?
Sometimes - lasting remission can follow treatment

IS IT AN EMERGENCY?
Sometimes - severe breathlessness or blue lips

LONG-TERM OUTLOOK
Often manageable, but serious breathing problems can occur

Overview

Pulmonary alveolar proteinosis, often shortened to PAP, is a rare lung condition. It causes a fatty, protein-rich substance called surfactant to collect inside the alveoli, the tiny air sacs where oxygen enters your blood.

Surfactant normally coats the air sacs and keeps them from collapsing. Old surfactant must be removed and recycled. In PAP, this clearing process does not work properly. The material builds up and makes it harder for oxygen to pass from the lungs into the bloodstream.

PAP is a group of related conditions rather than one single process. The most common form in adults is autoimmune PAP. In this form, the immune system makes antibodies that interfere with a signalling protein called granulocyte-macrophage colony-stimulating factor, or GM-CSF. GM-CSF helps immune cells in the lungs clear used surfactant.

Some people have mild or no symptoms. Others develop increasing breathlessness, low blood oxygen or lung infections. Identifying the type of PAP matters because treatment may involve removing material from the lungs, restoring GM-CSF signalling or treating another health condition.

Symptoms

Symptoms often develop gradually over weeks or months. They do not always match the amount of change seen on a lung scan. A person with extensive scan changes may have modest symptoms, while another person may be more limited.

Common symptoms include:

Low oxygen levels can cause headaches, poor concentration or a bluish or grey colour around the lips and fingertips. Clubbing, a rounded enlargement of the fingertips, may develop in long-standing cases but is not common.

Fever, worsening cough, chest pain or new mucus may indicate an infection rather than PAP alone. These symptoms need medical assessment because PAP can reduce the lungs' ability to clear certain germs.

Some people are diagnosed after an abnormal chest X-ray or CT scan taken for another reason.

- Shortness of breath, especially during activity
- A dry cough or a cough producing a small amount of mucus
- Tiredness and reduced stamina
- Difficulty keeping up with usual exercise or physical work
- Chest discomfort
- Unplanned weight loss

Causes and risk factors

Autoimmune PAP is the most common form in adults. The immune system produces antibodies against GM-CSF. Without effective GM-CSF signalling, cells called alveolar macrophages cannot remove surfactant normally. Autoimmune PAP is not caused by a lack of personal hygiene and does not spread between people.

Secondary PAP develops when another condition or exposure reduces the number or function

of these lung-cleaning cells. It can be associated with certain blood and bone marrow conditions, immune deficiencies, cancers or major exposure to mineral and industrial dusts. Treating the underlying problem may improve PAP, although this is not guaranteed.

Hereditary or congenital PAP results from a gene change affecting GM-CSF signalling, surfactant production or surfactant processing. Some forms begin in newborns or children. Others may not become apparent until later. The likely course and treatment differ according to the gene involved.

Smoking is common among adults diagnosed with PAP, but it is not the sole cause. People who have never smoked can develop the condition. Dust exposure may contribute in some cases, but many people with autoimmune PAP have no clear environmental trigger.

Diagnosis

Symptoms and imaging can suggest PAP, but they cannot confirm it by themselves. A high-resolution CT scan often shows hazy areas called ground-glass opacities. Thickened lines may cross these areas, creating a pattern known as "crazy paving." Other lung conditions can produce the same pattern.

Blood tests may measure oxygen levels and look for antibodies against GM-CSF. A strongly positive GM-CSF autoantibody test, together with compatible symptoms and imaging, supports autoimmune PAP. Additional blood tests may look for infection, blood cell abnormalities or an underlying condition.

Pulmonary function tests measure how well your lungs move air and transfer oxygen. PAP may reduce the lungs' ability to move gas into the bloodstream, even when the amount of air you can breathe in and out is close to normal.

Bronchoscopy may be used to confirm the diagnosis. During this procedure, a thin flexible camera is passed into the airways. Fluid is placed into a small part of the lung and collected for examination. In PAP, this bronchoalveolar lavage fluid often looks milky and contains characteristic surfactant material.

A lung biopsy is not usually needed when antibody testing, imaging and lavage findings are clear. It may be considered if the results are uncertain or another condition needs to be excluded. Genetic testing can help identify inherited forms, particularly in babies, children or families with more than one affected person.

Treatment and management

Treatment is based on the PAP type, symptoms, oxygen level and changes over time. People with mild, stable autoimmune PAP may be monitored without immediate treatment. Regular follow-up remains important because symptoms can progress.

Whole-lung lavage is an established treatment for people with significant breathing problems. It is performed under general anaesthesia at an experienced centre. One lung is ventilated while the other is repeatedly filled and drained with warmed salt water. This physically washes out accumulated surfactant. The other lung may be treated during a separate session.

Lavage can improve oxygen levels and activity tolerance, but it does not stop the immune process itself. Some people need only one course. Others need repeat procedures if material builds up again.

Inhaled GM-CSF treatment delivers the signalling protein directly to the lungs. It can improve surfactant clearance in autoimmune PAP and may be used as an initial treatment or after lavage. The exact product and treatment schedule differ between health systems and specialist centres.

If symptoms continue despite these treatments, specialists may consider therapies that reduce the harmful antibodies or remove them from the blood. These include medicines that act on antibody-producing immune cells and, in selected severe cases, plasma exchange. These options require specialist assessment because they have important risks and are not suitable for everyone.

Secondary PAP is managed by treating the underlying condition or reducing a relevant exposure when possible. Lung lavage may still be considered for significant breathing impairment. Treatment for inherited PAP is guided by the particular gene change and may involve specialised supportive care, lavage or, rarely, lung transplantation.

Oxygen can be used when blood oxygen is low. Antibiotics like amoxicillin or doxycycline treat confirmed or strongly suspected infections; they do not remove the surfactant buildup. Routine immune-suppressing treatment is generally avoided unless it is required for another condition, because suppressing immunity may increase the risk of infection.

Complications

PAP can lead to low blood oxygen and, in severe cases, respiratory failure. Respiratory failure means the lungs cannot provide enough oxygen or remove enough carbon dioxide to meet the body's needs.

The collected material also interferes with the lungs' immune defences. This increases susceptibility to infections, including infections caused by Nocardia , mycobacteria and certain

fungi. Some of these germs are uncommon in people with healthy lung defences.

Other possible complications include:

These complications do not affect everyone. Prompt assessment of new fever, chest pain or worsening respiratory symptoms can help identify infection or progression.

- Reduced ability to work, exercise or manage daily activities
- Repeated need for oxygen or lung-washing procedures
- Scarring of the lungs, known as pulmonary fibrosis
- Complications from severe or spreading infection
- Strain on the heart from long-term low oxygen

Prevention

Autoimmune PAP usually cannot be prevented because there is no established way to stop the GM-CSF antibodies from developing. Inherited PAP also cannot be prevented through lifestyle changes.

Some secondary cases may be linked to heavy exposure to silica, metal dust or other industrial particles. Following workplace controls, using suitable respiratory protection and avoiding unnecessary dust exposure may lower occupational lung risks, but it cannot prevent every case of PAP.

Stopping smoking and avoiding smoke or vaping aerosols will not cure PAP. However, doing so reduces additional irritation and damage to lungs that are already having difficulty exchanging oxygen.

If PAP is caused by an inherited gene change, genetic counselling can explain how that change may be passed through a family. It cannot predict every person's symptoms or course.

Living with it and what to expect

The course of PAP is different from person to person. Some people with autoimmune PAP remain stable for years or improve without treatment. Others have gradually worsening breathlessness or periods of improvement followed by recurrence.

Follow-up may include symptom reviews, oxygen measurements, pulmonary function tests and occasional imaging. These results help show whether the condition is stable and whether treatment is improving gas exchange. Routine follow-up is still useful when you feel well

because oxygen changes may appear before major symptoms.

After successful lavage or inhaled GM-CSF treatment, many people can increase their activity and return to work or usual routines. Improvement may be gradual. Treatment does not always return lung function to normal, and further treatment may be needed later.

Pacing can make daily tasks easier when stamina is limited. Pulmonary rehabilitation may help with safe exercise, breathing strategies and confidence during activity. If you use oxygen, your care team can explain how to use it safely at home and while travelling.

The outlook for secondary PAP is strongly influenced by the underlying condition. Inherited forms have a wide range of outcomes: some remain manageable, while severe forms beginning in infancy can be life-threatening. No single prediction applies to every type of PAP.

When to get medical help

Arrange routine medical review if you have gradually increasing breathlessness, a persistent cough, declining exercise tolerance or difficulty carrying out your usual activities. If you already have PAP, report a steady change rather than waiting for the next planned appointment.

Seek prompt same-day medical advice for:

Call emergency services for severe difficulty breathing, confusion, fainting, blue or grey lips, or an inability to speak in full sentences. These signs may indicate dangerously low oxygen or another serious problem.

- Fever with a new or worsening cough
- New mucus, especially if blood-stained
- Chest pain when breathing
- A clear drop in home oxygen readings from your usual range
- Breathlessness that is worsening faster than usual

Common questions

Is pulmonary alveolar proteinosis cancer?

No. PAP is not cancer. However, secondary PAP can occur alongside certain cancers or blood and bone marrow conditions. Tests may be needed to look for these underlying problems.

Is PAP contagious?

No. Autoimmune and inherited PAP cannot spread between people. A person with PAP can

develop a lung infection, but whether that infection is contagious depends on the germ involved.

Can pulmonary alveolar proteinosis go away on its own?

Sometimes autoimmune PAP improves without active treatment, especially when symptoms are mild. It can also remain stable or worsen. Monitoring helps detect changes before breathing becomes severely limited.

What does "crazy paving" on a CT scan mean?

Crazy paving describes hazy lung areas crossed by thickened lines, resembling irregular paving stones. It is common in PAP but is not specific to it. Infection, fluid in the lungs and other conditions can look similar.

Will I need my lungs washed?

Not everyone does. Whole-lung lavage is generally considered when PAP causes important symptoms, low oxygen or declining lung function. Mild, stable cases may only need monitoring, while some people are treated with inhaled GM-CSF.

Can I exercise with PAP?

Many people can exercise within limits set by their symptoms and oxygen level. Pulmonary rehabilitation can provide supervised guidance. Stop and seek medical advice if activity causes unusual chest pain, faintness or markedly worsening breathlessness.

Sources

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