CONDITION

Paroxysmal cold hemoglobinuria

Paroxysmal cold hemoglobinuria is an immune condition that suddenly destroys red blood cells, often after an infection.

WHAT IT IS
Cold-triggered autoimmune destruction of red blood cells

BODY SYSTEM
Blood and immune system

MAIN SYMPTOMS
Dark urine, fatigue, pallor, yellow skin

WHAT CAUSES IT
Immune reaction, often following infection

HOW IT IS DIAGNOSED
Blood tests and Donath-Landsteiner test

MAIN TREATMENT
Supportive care and treatment of underlying causes

CAN IT BE CURED?
Sometimes - acute cases usually resolve

IS IT AN EMERGENCY?
Sometimes - breathing trouble or fainting

LONG-TERM OUTLOOK
Usually full recovery after an acute episode

Overview

Paroxysmal cold hemoglobinuria, or PCH, is a type of autoimmune hemolytic anemia. "Autoimmune" means the immune system mistakenly attacks the body's own cells. "Hemolytic anemia" means red blood cells are destroyed faster than they can be replaced.

In PCH, an abnormal antibody attaches to red blood cells when blood becomes cooler, such as in the hands, feet, or other exposed areas. The antibody activates complement, a group of immune proteins. When the blood warms again, this immune reaction can break open the red blood cells inside the bloodstream.

The released red-cell pigment, called hemoglobin, may pass into the urine and turn it red, brown, or cola-colored. The loss of red blood cells can cause anemia, with tiredness, pallor, shortness of breath, or a fast heartbeat.

PCH is now most often seen as a sudden, short-lived condition in children after a viral illness. Adults can also develop it. Long-lasting or repeated PCH is less common and may be associated with another infection, an autoimmune condition, or a blood disorder.

Despite its name, attacks are not always clearly linked to cold exposure. "Paroxysmal" means sudden, and "hemoglobinuria" means hemoglobin in the urine.

Symptoms

Symptoms may appear suddenly, sometimes while you are recovering from a respiratory or other viral infection. They result from rapid red blood cell destruction and the anemia that follows.

Possible symptoms include:

Young children may be irritable, sleepy, less interested in food, or unwilling to play. Dark urine may be the first sign noticed by a parent or caregiver.

Urine color alone cannot confirm PCH. Blood in the urine, dehydration, liver problems, muscle injury, medicines, and some foods can also change urine color.

Symptoms can range from mild to severe. A rapid drop in red blood cells may cause marked weakness, breathlessness, fainting, or strain on the heart and circulation.

- Red, brown, or cola-colored urine
- Unusual tiredness or weakness
- Pale skin, lips, or nail beds
- Yellow skin or eyes, called jaundice
- Fever, chills, or feeling generally unwell
- Headache, dizziness, or light-headedness
- Shortness of breath or a fast heartbeat
- Pain in the back, abdomen, or legs

Causes and risk factors

PCH is caused by an antibody called the Donath-Landsteiner antibody. It is usually an IgG antibody, a type of immune protein. It commonly targets the P antigen, a marker found on the surface of most people's red blood cells.

The antibody acts in two temperatures. It attaches to red blood cells in cooler parts of the body and activates complement. As the blood returns to warmer areas, complement can rupture the cells. This is called intravascular hemolysis because the destruction occurs within blood vessels.

Modern cases are often associated with a recent infection, especially in children. The antibody may appear temporarily after the immune system responds to the infection. The exact infection is not always identified.

Less often, PCH may occur alongside:

Cold can trigger or worsen hemolysis while the antibody is present, but ordinary cold exposure does not by itself create the antibody. PCH is not usually inherited, and it is not passed directly from one person to another.

- Certain bacterial infections, historically including syphilis
- Autoimmune conditions
- Blood cancers or other disorders affecting immune cells
- No identifiable underlying condition

Diagnosis

Diagnosis begins with your symptoms, medical history, examination, and blood and urine tests. The tests look for anemia and evidence that red blood cells are breaking down.

A complete blood count measures the number of red blood cells and the level of hemoglobin, the protein that carries oxygen. A blood film allows the cells to be examined under a microscope.

Other tests may show:

The young cells are called reticulocytes. Their number can sometimes be low early in PCH, particularly during a severe episode, so a low result does not necessarily rule out hemolysis.

A direct antiglobulin test, also called a direct Coombs test, checks whether immune proteins are attached to red blood cells. In PCH, it is typically positive for complement but negative for IgG.

The Donath-Landsteiner test confirms the characteristic antibody. The blood sample must be collected, kept warm, and processed carefully because incorrect handling can cause a false-negative result. If suspicion remains high, the clinical team and laboratory may discuss repeating or adjusting the test.

Tests may also be used to look for a recent infection or another underlying condition. The choice depends on age, symptoms, examination findings, and whether hemolysis is persistent or recurrent.

- Increased bilirubin and lactate dehydrogenase, released during cell breakdown
- Low haptoglobin, a protein that binds free hemoglobin
- Hemoglobin in the urine
- An increased number of young red blood cells as the bone marrow responds

Treatment and management

Treatment depends on the severity of the anemia, how quickly red blood cells are being destroyed, and whether an underlying cause is found.

During active hemolysis, supportive care may include keeping the person and intravenous fluids warm, maintaining hydration, and closely monitoring blood counts, urine output, kidney function, and heart rate. Avoiding cold drinks and significant cold exposure may also reduce further antibody activity during the episode.

A blood transfusion may be needed if anemia is severe or causing concerning symptoms. Transfusion should not be delayed when it is clinically necessary. The transfusion service can select compatible blood and use appropriate warming procedures.

Any identified underlying infection or other condition is treated on its own merits. Antibiotics like amoxicillin or doxycycline do not treat PCH itself and are only useful when there is a bacterial infection that requires them.

Corticosteroids like prednisone or hydrocortisone are sometimes started before the exact type of autoimmune hemolytic anemia is known. Their benefit in typical acute PCH is uncertain, and many cases recover with supportive care alone. Decisions about continuing immune-suppressing treatment are made after the diagnosis and severity are reviewed.

For rare, severe, persistent, or repeatedly recurring cases, a hematology specialist may consider treatments that affect antibody-producing cells or the complement system. Evidence for these approaches in PCH is limited, so they are not routine treatment for most people.

Complications

The main complication is severe anemia. If the red blood cell count falls quickly, the body may have difficulty delivering enough oxygen to tissues. This can cause fainting, breathing

difficulty, chest discomfort, or cardiovascular strain.

Large amounts of free hemoglobin can affect the kidneys. Dehydration may increase this risk. Urine output and kidney blood tests are therefore monitored during a significant episode.

Other possible complications include:

Most acute childhood cases improve without lasting organ damage. Serious complications are more likely when hemolysis is rapid, anemia is profound, or another major health condition is present.

- A need for urgent blood transfusion
- Temporary bone marrow difficulty replacing lost red blood cells
- Ongoing hemolysis if the antibody remains active
- Complications from an underlying infection or blood disorder

Prevention

PCH usually cannot be prevented. The abnormal antibody often develops unexpectedly after an infection, and there is no established way to stop this immune response from occurring.

While hemolysis is active, staying warm and avoiding major cold exposure may reduce further antibody binding. This is a short-term management measure, not a guarantee against another episode.

Routine steps that reduce the spread of infections, such as handwashing and recommended vaccination, support general health but cannot reliably prevent PCH. Because the condition is not usually inherited, family members do not normally require screening.

Living with it and what to expect

Acute PCH often begins suddenly and then settles as the temporary antibody disappears. Hemolysis may stop over days, while hemoglobin levels and energy can take longer to return to normal. Follow-up blood tests help show whether red blood cells are recovering and whether signs of hemolysis have cleared.

During recovery, tiredness may continue even after the urine returns to its normal color. Activity can usually be increased gradually according to symptoms and medical advice. Significant cold exposure may need to be avoided until the episode has resolved.

Most children with infection-associated PCH have a single episode and recover fully.

Recurrence is possible but is not the usual course. Long-lasting PCH is more likely when there is a continuing infection, autoimmune condition, or blood disorder.

Routine follow-up may include repeat blood counts, markers of hemolysis, and kidney tests. Persistent anemia, repeated dark urine, or a return of jaundice needs reassessment rather than waiting for the next planned appointment.

PCH itself does not usually require lifelong treatment after an isolated episode. If it is linked to another condition, longer-term care will be shaped by that condition.

When to get medical help

Seek prompt medical assessment for unexplained red or brown urine, yellow skin or eyes, marked pallor, or a sudden loss of energy. These symptoms have several possible causes and require testing.

If you have already been diagnosed with PCH, contact your healthcare team promptly if dark urine returns, tiredness worsens, fever persists, or you are passing less urine than usual. These are reasons for earlier review, not just routine follow-up.

Seek emergency care for:

For a baby or young child, emergency signs also include unusual limpness, difficulty waking, struggling to breathe, or being too weak to drink.

- Difficulty breathing at rest
- Chest pain
- Fainting or inability to stay awake
- Confusion or severe weakness
- A very fast heartbeat with dizziness
- Very little or no urine
- Rapidly worsening pallor or jaundice

Common questions

Is paroxysmal cold hemoglobinuria a type of cancer?

No. PCH is an immune-mediated blood condition, not a cancer. In uncommon cases, especially in adults, it can occur alongside a blood cancer or another disorder. Finding PCH does not mean that cancer is present.

Is PCH contagious?

PCH itself is not contagious. A viral or bacterial infection that came before it may be contagious, but the Donath-Landsteiner antibody and red blood cell destruction cannot spread to someone else.

Does cold weather always trigger an attack?

No. The antibody reacts more readily at cooler temperatures, but many episodes follow an infection without an obvious cold trigger. Cold avoidance is helpful during active hemolysis, but it does not replace medical monitoring.

Will I or my child need a blood transfusion?

Not everyone does. Transfusion is considered when anemia is severe, falling quickly, or causing symptoms such as breathing difficulty, fainting, or cardiovascular strain. The decision is based on the whole clinical picture, not one blood count alone.

Does PCH go away completely?

Often, especially when it occurs in a child after an infection. The antibody may disappear and the blood count can return to normal. Persistent or recurrent PCH is possible and usually prompts investigation for an underlying condition.

Can PCH come back after recovery?

It can, but repeated episodes are less typical than a single acute episode. New dark urine, jaundice, pallor, or unusual weakness should be assessed even if previous follow-up tests were normal.

Sources

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