CONDITION

Hyperkalemic periodic paralysis

Hyperkalemic periodic paralysis is an inherited condition that causes temporary attacks of muscle weakness or paralysis.

WHAT IT IS
Inherited condition causing episodes of muscle weakness

BODY SYSTEM
Nervous system and skeletal muscles

MAIN SYMPTOMS
Sudden weakness, temporary paralysis, stiffness, muscle aches

WHAT CAUSES IT
Inherited sodium channel gene changes

HOW IT IS DIAGNOSED
Genetic testing and specialized muscle electrical testing

MAIN TREATMENT
Trigger management and medicines that reduce attacks

CAN IT BE CURED?
No - the inherited gene change remains

IS IT AN EMERGENCY?
Sometimes - if breathing, swallowing, or heartbeat is affected

LONG-TERM OUTLOOK
Normal lifespan for most; fixed weakness can develop

Overview

Hyperkalemic periodic paralysis is a rare inherited condition that causes sudden episodes of muscle weakness. During a severe episode, affected muscles may be temporarily unable to move. This is called paralysis.

The condition affects sodium channels in skeletal muscle. These channels help muscle cells respond to nerve signals. When they do not work normally, muscle cells can become temporarily unable to contract.

"Hyperkalemic" means that the potassium level in the blood may be high during an attack. However, potassium can also be within the normal range. A normal result during an attack does not automatically rule out the condition.

Attacks often begin in infancy or childhood. They may occur after resting following exercise, on waking, during fasting, or after eating a large amount of potassium-rich food. Episodes are usually temporary, but some people develop lasting muscle weakness later in life.

Hyperkalemic periodic paralysis is not the same as ordinary hyperkalemia caused by kidney problems, medicines, or another medical condition. Those causes of high potassium need separate assessment because significant hyperkalemia can affect the heart.

Symptoms

The main symptom is an attack of muscle weakness that comes and goes. An attack may affect the arms, legs, shoulders, hips, or trunk. The legs are often affected more than the arms.

Symptoms may include:

Attacks often begin after a trigger. Weakness may develop while resting after physical activity rather than during the activity itself. Some people notice symptoms when they wake in the morning.

An episode may last from several minutes to a few hours. Frequency differs widely. Some people have attacks most days, while others have them only occasionally.

Sensation and awareness usually remain normal. Numbness, confusion, loss of consciousness, or weakness affecting only one side of the body is not typical and should not automatically be assumed to be a periodic paralysis attack.

Some people also have myotonia , meaning a muscle is slow to relax after contracting. For example, the hand may briefly remain stiff after gripping something. Cold temperatures can make stiffness worse in some people.

Breathing and swallowing muscles are usually spared, but they can rarely be affected. Weakness involving these muscles needs urgent medical attention.

- Sudden heaviness or weakness in several muscles
- Temporary inability to move affected muscles
- Muscle stiffness, especially before or between attacks
- Muscle aching or tiredness after an episode
- Eyelid stiffness or difficulty opening the eyes after closing them tightly

Causes and risk factors

Hyperkalemic periodic paralysis is most often caused by a change in the SCN4A gene. This gene provides instructions for a sodium channel in skeletal muscle.

The altered channel may stay open longer than it should. This disrupts the muscle cell's electrical balance and can make the muscle temporarily unable to contract.

The condition is usually inherited in an autosomal dominant pattern. This means a person with the gene change has a 1 in 2 chance of passing it to each child. Symptoms can still differ greatly among members of the same family.

A person may have no known family history. This can happen when relatives had mild or unrecognized symptoms, or when the gene change arose for the first time in that person.

Triggers do not cause the inherited condition, but they may bring on attacks. Commonly reported triggers include:

Not everyone has the same triggers. Keeping a record of attacks can help identify patterns without making unnecessary changes to diet or activity.

- Rest after strenuous or unaccustomed exercise
- Going without food for a long time
- Potassium supplements or a large potassium load
- Cold temperatures
- Physical or emotional stress
- Illness
- Certain medicines or medical procedures

Diagnosis

Diagnosis usually begins with the pattern of attacks, age at which they started, possible triggers, and family history. A neurological examination may be normal between episodes.

Blood tests taken during an attack can measure potassium and look for other causes of weakness. Potassium may be mildly raised or normal in hyperkalemic periodic paralysis, so this result cannot confirm or exclude the condition by itself.

An electrocardiogram, or ECG, may be used to check the heart's electrical activity when potassium is elevated. Other tests may assess kidney function, thyroid function, blood glucose, and muscle enzymes. These help rule out acquired causes of paralysis or abnormal potassium.

Electromyography , usually called EMG, measures electrical activity in muscles. A specialized long exercise test may show a characteristic change in muscle response after exercise.

Genetic testing can identify a disease-causing change in SCN4A and confirm the diagnosis in many people. A negative genetic test does not settle every case, so results are considered alongside symptoms and electrical testing.

Deliberately giving potassium to trigger an attack can be dangerous and is not routinely needed. It should never be attempted at home.

Treatment and management

Treatment aims to reduce how often attacks occur, limit their severity, and protect long-term muscle function. The approach is based on each person's attack pattern and test results.

A management plan may include:

Extreme restriction of dietary potassium is not appropriate for everyone and can make a balanced diet difficult. A clinician or dietitian familiar with the condition can help identify whether particular foods or meal patterns are relevant.

Medicines used to prevent attacks include carbonic anhydrase inhibitors, such as dichlorphenamide or acetazolamide, and sometimes certain diuretics like furosemide or hydrochlorothiazide . Diuretics change how the kidneys handle salt and water. Response differs, and acetazolamide can worsen symptoms in some people with particular SCN4A changes.

Myotonia may require separate treatment. Any medicine used for this condition needs monitoring for side effects and changes in blood electrolytes, including potassium.

During an attack, mild activity may help some people if it can be done safely. Carbohydrate may also help shift potassium into cells in selected situations, but an individualized attack plan is important.

Severe weakness or significantly elevated potassium may require treatment in an emergency department. Hospital treatment may include heart monitoring and medicines that lower potassium or protect the heart. Do not take extra medicine or potassium-lowering treatment unless it is part of an agreed plan.

Potassium supplements, potassium-containing salt substitutes, and medicines that raise potassium may need review. Do not stop prescribed medicines without medical advice.

- Recognizing and reducing personal triggers
- Eating regular meals and avoiding prolonged fasting
- Cooling down gradually after strenuous activity

- Using gentle movement at the first sign of some attacks
- Reviewing medicines and supplements that affect potassium
- Taking preventive medicine when attacks remain troublesome

Complications

Temporary weakness can lead to falls, injuries, or difficulty completing daily activities. Frequent attacks may also disrupt school, work, sleep, and exercise.

Some people gradually develop fixed weakness , meaning weakness that remains between attacks. This often affects muscles around the hips or shoulders and becomes more noticeable later in adulthood. It does not occur in everyone.

Markedly high potassium can disturb the heart's rhythm. This is not the usual feature of a mild attack, but palpitations, fainting, chest pain, or an abnormal ECG require urgent assessment.

Preventive medicines can also cause problems. Depending on the medicine, these may include tingling, tiredness, changes in thinking, dehydration, low potassium, or kidney stones. Regular follow-up may include blood and urine tests.

Prevention

The inherited gene change cannot currently be prevented or removed. There is no known way to guarantee that attacks will stop.

Many attacks can still be reduced by learning personal triggers, avoiding prolonged fasting, cooling down after exercise, and following an agreed treatment plan. A symptom diary can record activity, meals, illness, temperature, medicines, and attack length.

Genetic counselling can explain inheritance, testing for relatives, and reproductive options. Testing is a personal choice, particularly for relatives who have no symptoms.

Before surgery or a procedure requiring anesthesia, tell the anesthesia team about the condition. Fasting, temperature changes, stress, and some medicines can affect muscle function or potassium. Advance planning allows these factors to be monitored.

Living with it and what to expect

Hyperkalemic periodic paralysis is lifelong, but its pattern can change. Attacks often begin

early in life and may become less frequent in later adulthood. This is not guaranteed, and some people develop more persistent weakness even as temporary attacks decrease.

Many people remain active between episodes and have a normal lifespan. Work, education, travel, and exercise are usually possible with planning. The main practical challenge is often the unpredictable timing of weakness.

Regular, moderate activity may help maintain strength and general health. Sudden intense exercise followed by complete rest is more likely to trigger symptoms than steady activity with a gradual cool-down.

A written attack plan can explain what usually helps, which symptoms are unusual, and when emergency care is needed. A medical alert card or bracelet may be useful, especially if severe attacks occur.

Routine follow-up is used to review attack frequency, strength between episodes, medication effects, potassium levels, and kidney health. New persistent weakness should be assessed rather than treated as an expected part of aging.

When to get medical help

Seek emergency medical help if weakness:

A laboratory result showing significantly elevated potassium also needs prompt medical assessment, even if symptoms seem mild. High potassium can have causes other than periodic paralysis and may affect the heart without obvious warning symptoms.

Arrange routine medical follow-up if attacks are becoming more frequent, lasting longer, or changing in character. Also seek review for new weakness between attacks, repeated falls, medication side effects, or difficulty managing triggers.

A first unexplained episode of paralysis needs medical assessment. Several conditions can cause sudden weakness, and the correct treatment depends on the cause.

- Affects breathing, swallowing, or the ability to hold up the head
- Occurs with chest pain, a racing or irregular heartbeat, or fainting
- Is severe enough to prevent safe movement
- Affects one side of the body or occurs with facial drooping or speech difficulty
- Comes with confusion, loss of consciousness, or a serious injury

Common questions

Is potassium always high during an attack?

No. Potassium may be mildly elevated, may rise compared with your usual level, or may remain within the laboratory's normal range. Diagnosis does not rely on one potassium result.

Is this the same as hypokalemic periodic paralysis?

No. Both cause attacks of weakness, but they usually involve different potassium patterns, triggers, and gene changes. Treatment that helps one form may not be suitable for another.

Can I exercise if I have hyperkalemic periodic paralysis?

Many people can exercise. Steady, moderate activity with a gradual warm-up and cool-down is often better tolerated than sudden intense exertion followed by complete rest. An activity plan should account for your usual triggers and baseline strength.

Do I need to avoid all high-potassium foods?

Usually not. Extreme dietary restriction can cause nutritional problems. Some people react to large potassium loads or particular foods, while others do not. A symptom diary and professional dietary advice can help guide changes.

Will my children inherit it?

When a person has an autosomal dominant SCN4A change, each child has a 1 in 2 chance of inheriting that change. Inheriting it does not predict exactly how mild or severe symptoms will be.

Can hyperkalemic periodic paralysis be cured?

There is currently no cure that removes the inherited gene change. Trigger management and preventive medicines can reduce attacks for many people, while monitoring can identify persistent weakness or treatment side effects.

Sources

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