CONDITION

Idiopathic basal ganglia calcification

Idiopathic basal ganglia calcification is calcium buildup in deep brain areas without an identified medical cause.

WHAT IT IS
Calcium deposits in deep areas of the brain

BODY SYSTEM
Nervous system

MAIN SYMPTOMS
Movement problems, thinking changes, mood changes, seizures

WHAT CAUSES IT
Usually inherited or unknown

HOW IT IS DIAGNOSED
Brain CT and tests excluding secondary causes

MAIN TREATMENT
Symptom-based medicines, therapy, and supportive care

CAN IT BE CURED?
No - deposits cannot currently be removed

IS IT AN EMERGENCY?
Sometimes - sudden stroke-like symptoms or prolonged seizure

LONG-TERM OUTLOOK
Many remain stable; some develop progressive neurological symptoms

Overview

Idiopathic basal ganglia calcification is a condition in which calcium deposits collect in deep parts of the brain. The basal ganglia help control movement, coordination, learning, emotions, and some aspects of thinking.

"Idiopathic" means that no separate medical cause has been found. Some cases are caused by an inherited gene change. These may be called primary familial brain calcification. Other cases occur without a known family history or identifiable gene change.

The older term "Fahr disease" is still used online and in some medical records. However, its meaning has not always been consistent. More specific terms, such as primary familial brain calcification or idiopathic basal ganglia calcification, are generally clearer.

The deposits are usually present on both sides of the brain. They may also affect areas beyond the basal ganglia, including the thalamus, cerebellum, and the brain's white matter.

Some people have no symptoms and learn about the calcification after a brain scan for another reason. Others develop movement, psychiatric, cognitive, or seizure-related symptoms. The amount of calcification seen on a scan does not reliably predict how a person will feel or function.

Small areas of brain calcification can occur with ageing and do not always mean that someone has this condition. A diagnosis requires the scan pattern to be considered alongside symptoms, medical history, and tests for other causes.

Symptoms

Symptoms differ widely, even among relatives with the same genetic change. They may begin in adulthood, although the age of onset is not predictable. Some people remain free of symptoms.

Movement symptoms can include:

"Parkinsonism" describes a group of movement problems, such as slowness and stiffness. It does not necessarily mean that a person has Parkinson's disease.

Changes in thinking or behavior may include difficulty with memory, attention, planning, or judgment. Some people experience depression, anxiety, irritability, loss of motivation, hallucinations, or other changes in behavior. These symptoms can have many possible causes and need careful assessment rather than being assumed to come from the calcification.

Other possible symptoms include seizures, headaches, dizziness, urinary control problems, or changes in speech. Seizures are sudden bursts of abnormal electrical activity in the brain. They can cause shaking, loss of awareness, unusual sensations, or brief periods of staring.

Symptoms may be mild, intermittent, or slowly progressive. A person can also have substantial calcification on a scan without noticeable neurological problems.

- Slowness, stiffness, or shaking similar to parkinsonism
- Involuntary twisting, repetitive movements, or muscle spasms
- Unsteady walking, poor balance, or frequent falls
- Slurred speech or difficulty coordinating swallowing

Causes and risk factors

Idiopathic basal ganglia calcification is considered a primary brain calcification condition. This means the deposits are not explained by another illness, medicine, toxin, infection, or injury.

Several genes have been linked to primary familial brain calcification. These genes affect processes such as phosphate transport, the health of small blood vessels, or the brain's protective barrier between blood and tissue. Changes in these processes may allow calcium and phosphate deposits to build up around small blood vessels.

Some forms follow an autosomal dominant inheritance pattern. This means a person can inherit the gene change from one parent. Other forms are autosomal recessive, meaning a person usually inherits one altered copy from each parent. Having a gene change does not predict the exact symptoms, age of onset, or degree of disability.

A family history may increase the chance of an inherited form, but its absence does not rule one out. Relatives may have mild symptoms, no symptoms, or no brain imaging. A new gene change can also occur in someone without an affected parent.

Before calling the condition idiopathic or primary, clinicians look for secondary causes. These include disorders of the parathyroid glands, abnormal calcium or phosphate levels, certain infections, mitochondrial conditions, toxic exposures, and some other neurological disorders. When another cause is found, the condition is classified and managed differently.

Diagnosis

A brain CT scan is the main imaging test used to identify and map calcification. CT uses X-rays and shows calcium more clearly than MRI in most cases. The usual pattern involves both basal ganglia and may include other brain regions.

A scan alone does not confirm that the condition is idiopathic. Diagnosis also involves checking whether the pattern could be age-related or caused by another medical problem.

The assessment may include:

Genetic testing may identify a known cause, particularly when several relatives are affected or the scan pattern is typical. A positive result can clarify inheritance and may help relatives make informed choices about testing. A negative result does not exclude the condition because not every genetic cause is known or detectable.

Genetic counselling can help explain what a result means for you and your biological relatives. Testing an adult without symptoms is a personal decision because a positive result cannot show when symptoms will start or whether they will develop.

- A medical and family history
- A neurological examination of movement, balance, speech, and coordination
- Assessment of memory, thinking, mood, or behavior when relevant
- Blood tests for calcium, phosphate, magnesium, kidney function, parathyroid hormone, and related measures
- Additional tests guided by symptoms, history, or possible exposures

Treatment and management

There is currently no treatment that removes the brain deposits or reliably stops them from increasing. Care focuses on symptoms, safety, independence, and quality of life.

Treatment is tailored to the problems a person has. It may include medicines for seizures, depression, anxiety, psychosis, involuntary movements, or parkinsonism. Responses can be mixed, so medicines may need review over time. Treatment decisions also consider possible side effects, especially when balance, swallowing, alertness, or thinking are already affected.

Physical therapy can support strength, walking, balance, and fall prevention. Occupational therapy can help with daily tasks, work, home adaptations, and mobility equipment. Speech and language therapy may help with communication or swallowing.

If swallowing becomes difficult, an assessment can identify safer food textures and eating techniques. Nutrition support may be useful when eating takes longer or weight is being lost.

Mental health symptoms deserve the same attention as movement symptoms. Psychological therapy, social support, and appropriate medicines may all have a role.

Routine follow-up is based on symptoms rather than the scan alone. It may include neurological review, medicine monitoring, falls assessment, cognitive testing, or support for work and daily activities. Repeated CT scans are not always useful because changes on imaging may not match changes in symptoms.

Complications

Not everyone develops complications. When symptoms are present, possible problems include falls, injuries, reduced mobility, loss of independence, and difficulty continuing work or driving.

Swallowing problems can lead to weight loss, dehydration, or food and liquid entering the

airway. This is called aspiration and can sometimes cause a chest infection.

Seizures can cause injuries or temporary restrictions on driving and certain activities. Local driving rules apply after a seizure or when movement, vision, judgment, or alertness is impaired.

Changes in memory, judgment, mood, or behavior may affect relationships, finances, medicine management, and personal safety. Severe depression, psychosis, or cognitive decline can place additional pressure on both the affected person and those supporting them.

These complications are possibilities, not an expected outcome for everyone.

Prevention

Idiopathic basal ganglia calcification usually cannot be prevented. In inherited forms, the gene change is present from conception. Lifestyle changes have not been shown to prevent the brain deposits.

This is different from secondary brain calcification caused by another medical condition. Finding and treating abnormal calcium or parathyroid hormone levels may prevent further health problems, but it does not necessarily remove deposits that are already present.

If a genetic cause is identified, genetic counselling can explain inheritance and family-planning options. This supports informed choices but does not change the condition in someone who already has it.

General measures such as regular activity, adequate sleep, limiting alcohol, and not smoking can support overall health. They should not be presented as ways to prevent this specific condition.

Living with it and what to expect

The long-term course is difficult to predict. Some people remain symptom-free or have mild, stable symptoms for years. Others develop gradually increasing movement, cognitive, psychiatric, or seizure-related problems.

Calcification may become more extensive over time, but scan changes do not closely match day-to-day ability. A person with widespread deposits may function well, while someone with less visible calcification may have troublesome symptoms.

Regular review is most useful when it focuses on practical changes: walking, falls, swallowing, seizures, mood, memory, work, driving, and daily independence. New symptoms should not

automatically be blamed on the calcification because they may have another treatable cause.

Planning can help if symptoms progress. This may involve home adjustments, workplace support, advance care discussions, or identifying trusted people who can help with finances and medical decisions. Such planning does not mean that major disability is certain.

Family members can have very different experiences, even when they share the same gene change. Genetic results and brain scans cannot give a precise personal forecast.

When to get medical help

Arrange a routine medical appointment if you develop persistent changes in movement, balance, memory, mood, behavior, or speech. Review is also appropriate after an unexplained fall, a possible seizure, or new swallowing difficulty.

If you already have the condition, seek prompt advice when symptoms begin affecting eating, walking, work, driving, medicine use, or daily safety. Gradual changes usually need planned assessment rather than emergency care.

Seek emergency help for:

Sudden stroke-like symptoms should not be assumed to come from existing calcification. They need emergency assessment.

- Sudden facial drooping, arm weakness, speech difficulty, or loss of coordination
- A first seizure, a seizure lasting several minutes, or repeated seizures without recovery
- Severe breathing difficulty or choking that does not clear
- Sudden severe confusion, loss of consciousness, or a serious fall with head injury
- Thoughts of suicide or an immediate risk of harm

Common questions

Is idiopathic basal ganglia calcification the same as Fahr disease?

"Fahr disease" is an older and less precise term often used for primary or familial brain calcification. Idiopathic basal ganglia calcification describes the scan finding and the absence of an identified secondary medical cause.

Does calcium in the brain come from eating too much calcium?

No. Dietary calcium has not been shown to cause this condition. Do not restrict calcium without medical advice, since too little calcium can harm bones and other body functions.

Does the condition always cause dementia?

No. Some people have no cognitive symptoms. Others develop problems with memory, attention, planning, or judgment, and a smaller group may develop substantial cognitive impairment. A scan cannot predict this with certainty.

Can the calcium deposits be removed?

There is no established treatment that safely removes these deposits from brain tissue. Surgery is not used to clear them. Treatment instead targets seizures, movement problems, mental health symptoms, and practical difficulties.

Will my children inherit it?

That depends on whether a genetic cause is present and how it is inherited. Some forms can be passed from one affected parent, while others require altered gene copies from both parents. Genetic testing and counselling can clarify the chance for a particular family.

Can I still work, exercise, or drive?

Many people continue their usual activities, especially when they have no symptoms or only mild symptoms. Safety may need review if you have seizures, falls, slowed reactions, impaired judgment, or vision problems. Driving requirements are set by local licensing authorities.

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