

Corticobasal degeneration (CBD) is a rare progressive neurodegenerative disease that causes parts of the brain to deteriorate over time. As a result of this deterioration, people living with CBD experience increased difficulty with their coordination, movement, thinking and speech.

About corticobasal degeneration

CBD is a rare form of dementia that is caused by an overproduction of a protein in the brain called tau. This build-up of tau protein causes areas of the brain to become increasingly damaged and to shrink over time. The parts of the brain that are most affected by CBD are the outer “thinking” layer of the brain, known as the cortex, and the deeper areas of the brain, called the basal ganglia, which helps with movement.

As the cortex is responsible for higher level cognitive functioning like thinking and understanding, and the basal ganglia helps people to perform smooth movements, CBD may affect both the physical and cognitive functioning of people with the disease.

Other names used for CBD include: Corticobasal Ganglionic Degeneration, Corticobasal Syndrome and Cortical-basal Ganglionic Degeneration.

What are the symptoms?

Symptoms of CBD are most commonly seen in people between the ages of 60 and 80, but may begin in some people in their 40's. CBD can be confused with Parkinson's disease and Progressive Supranuclear Palsy because some symptoms, like stiffness or rigid movements, can resemble some of the common characteristics of these other neurodegenerative diseases.

Early symptoms of this disease are often related to difficulties with the person's movement or can present as changes with planning, multitasking, speech, language, or praxis (practice). A person may only experience the movement symptoms in one of their hands, arms or legs. It is also not uncommon for the early physical symptoms to appear only on one side of the body. As the disease progresses, both sides of the body will become affected.

Similar to symptoms of Parkinson's disease, people with CBD can experience movement difficulties such as rigidity, lack of coordination, and limb dystonia (involuntary tightening of the affected limb). People with CBD may complain that a limb affected by the disease does not feel like part of their body. This is known as the “alien limb” sensation. Unlike people with Parkinson's disease, tremors have been found to be less common in those who are living with CBD.

While it may not be a common early symptom of CBD, changes in cognitive abilities may be experienced as the disease progresses. People with the cognitive symptoms of CBD typically experience difficulties with recalling words, simple calculations, carrying out a plan, praxis and slow or slurred speech. Visual spatial disorientation can be seen as a feature of this disease.

How is corticobasal degeneration diagnosed?

It can be challenging to diagnose CBD early in the progression of the disease as there are a number of symptoms that are shared with similar neurodegenerative diseases, including Parkinson's disease, Alzheimer's disease, frontotemporal dementia, Lewy body dementia, progressive supranuclear palsy and multiple system atrophy.

People who show signs of cognitive difficulties prior to movement symptoms may be misdiagnosed with Alzheimer's disease or frontotemporal dementia. It is usually not until movement symptoms develop that CBD is considered. CBD may take several months or even years to diagnose. Making a diagnosis of CBD becomes easier for physicians when the person presents with both cognitive and physical symptoms.

There is no single test to diagnose CBD. Physicians assess patterns and types of symptoms to help rule out other conditions that could be causing the symptoms, such as Parkinson's disease, stroke or motor neuron disease. Brain imaging (MRI) is often used to help detect any abnormal changes in the brain that are features of CBD, such as the shrinkage of the brain's cortex. A thorough assessment may also include neuropsychological testing to evaluate the impact of symptoms on the person's mental abilities.

What are the causes or the risk factors?

At present, there is no known cause of CBD and common risk factors have not been identified. However, similar to other types of dementia, CBD is characterized by an excess amount of the tau protein in the brain. It is still unknown what causes this surplus of the tau protein, and why it results in the deterioration of brain cells.

Is there treatment?

Currently, there is no known cure for CBD. Medications that are often used to manage symptoms of Parkinson's disease can be tried in CBD but are usually not as effective. Cholinesterase inhibitors used to improve alertness and cognition in people with Alzheimer's disease do benefit some people with CBD.

Therapeutic approaches can support people living with CBD to manage symptoms of their disease. Occupational and physical therapies that provide passive range of motion may help to maintain physical functioning throughout the progression of the disease. Early speech therapy has also been found to help delay difficulties with verbal communication.

Support is available:

Visit the Alzheimer Society's website at www.alzheimer.ca or contact your local Alzheimer Society.

Additional resources:

- Alzheimer's Society UK:
http://www.alzheimers.org.uk/site/scripts/documents_info.php?documentID=135
- Cure PSP:
<http://www.psp.org/education/cbd.html>

- Parkinson Society Canada:
http://www.parkinson.ca/site/c.kgLNiWODKpF/b.8649607/k.49F7/Corticobasalganglionic_Degeneration.htm
- The Association for Frontotemporal Degeneration:
<http://www.theaftd.org/frontotemporal-degeneration/disorders/corticobasal-degeneration>

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