

## CORTICOBASAL DEGENERATION (CBD)

---

Corticobasal degeneration, sometimes referred to as corticobasal ganglionic degeneration (CBGD), is a heterogeneous disease which clinically, genetically and pathologically is similar to, or overlaps with frontotemporal dementia (FTD). For this reason, CBD is considered to be part of the 'Pick complex' of neurodegenerative diseases.

CBD was first described in 1968 by Rebeiz and colleagues, who immediately recognized its potential relationship to FTD based on macroscopic and microscopic analyses of CBD brains. Historically, CBD patients have been diagnosed on the basis of movement problems which sometimes appear similar to Parkinson's disease (PD). Unlike PD, however, CBD patients typically do not respond significantly to PD medicines, such as levodopa/carbidopa (Sinemet). Also, many symptoms of CBD are not found in PD patients. For this reason CBD is often referred to as a 'Parkinson's-plus' syndrome.

When a diagnosis of CBD is suspected, it is important to refer the patient to a neurologist who is experienced with this disorder. This is because the constellation of symptoms and problems experienced by affected individuals and their caregivers is unique. There have been significant advances in the understanding of CBD over the past 10 years, and as a result, improved counseling, support and symptomatic treatments are now available. We are actively involved in research to better understand the pathophysiology of CBD.

### Demographics

CBD typically occurs in patients between 45 and 70. In our experience, women are affected more commonly than men. Rarely, there is a family history of dementia, psychiatric problems or a movement disorder.

### Symptoms

Patients with CBD present with either a movement disorder or cognitive deficits. As the disease progresses, most patients will eventually develop both types of symptoms, often with a delay of 2-3 years.

### Movement Problems in CBD

A characteristic feature of movement symptoms in CBD is striking asymmetry of involvement. Most frequently symptoms begin insidiously in one hand or arm, less commonly in one leg. Rarely, symptoms may involve the mouth and facial muscles.

Many patients will complain initially of a subtle change in sensation or an inability to make the affected limb follow commands. This latter deficit is called **apraxia** and may be

confused for clumsiness or weakness. There may be difficulties in completing specific tasks, such as opening a door or brushing one's teeth or using tools, such as a can opener. When a leg is affected initially, a patient may have problems with complex movements such as dancing; or when more severe, a patient may begin to trip and fall. Some patients will experience an involuntary stiffening, twisting or contraction of the affected limb called **dystonia**. There may be uncontrolled jumping of the limb when it is tapped gently or when the patient is startled, called **myoclonus**.

Finally, CBD patients often complain that the affected limb feels like it is not a part of their body, a sensation called **alien limb**. Sometimes an alien limb will move on its own, in an uncontrollable way. For example, an alien hand will rise to touch the patient's face. Alien limb phenomenon was dramatized by the actor Peter Sellers in the film **Dr. Strangelove**.

Movement symptoms tend to progress slowly from one side of the body to the other, or from leg to arm on the same side of the body.

### **Cognitive Problems in CBD**

Patients with CBD who present with cognitive difficulties are usually initially diagnosed with frontotemporal dementia or Alzheimer's disease. It is only after they develop movement symptoms that the diagnosis of CBD is entertained. Occasionally, a diagnosis of CBD is not apparent until a patient's brain is examined at autopsy.

Progressive difficulty with language is a common cognitive complaint in CBD. This most commonly involves difficulty with expression of language, such as word finding difficulty or naming problems. Reading, writing and simple mathematical calculations may also be impaired.

Personality changes, inappropriate behavior, repetitive and/or compulsive activities similar to those seen in FTD (see FTD description) are also common in CBD. Short-term memory problems, such as repeating questions or misplacing objects are also common.

Many patients with the movement difficulties of CBD will also have mild cognitive problems when they are evaluated in a specialized dementia clinic.

### **Treatment**

At this time, there is no specific treatment for CBD. Instead individual symptoms are targeted with specific medications. For example, rigidity and difficulty walking may partially respond to treatments for Parkinson's disease. Dystonia and myoclonus may respond to muscle relaxants or anti-seizure medications. Memory and behavior problems may respond to treatments for Alzheimer's disease and/or depression.