Corticobasal Degeneration

Dementia Neuropsychology · https://neuro.vip/neuropsychology/dementia-neuropsychology/corticobasal-degeneration/

Corticobasal Degeneration (CBD) is a rare, **progressive neurodegenerative disease** that presents with a **distinct combination of movement, cognitive, and behavioural symptoms**. Given its heterogeneous presentation and **overlap with other neurodegenerative disorder**s, accurate clinical identification and neuropsychological assessment are essential.

CBD typically develops in **adults over the age of 60** and has a **steadily progressive course**, often leading to **severe disability and death within 6–8 years** of symptom onset. Originally thought to be primarily a movement disorder, there is now strong recognition of cognitive and behavioural changes, sometimes even preceding motor symptoms. This makes the involvement of clinical neuropsychologists vital, both for diagnosis and management.

CBD is considered a tauopathy, marked by **abnormal tau protein deposits** affecting the cortex (especially **frontoparietal regions**) and **basal ganglia**.

Clinical Features of Corticobasal Degeneration

Motor Symptoms

CBD's motor manifestations are often **strikingly asymmetric**—symptoms usually affect one side of the body significantly more than the other. Key motor features include:

- **Limb rigidity and akinesia** (stiffness and slowness)
- **Dystonia** (abnormal sustained postures)
- **Myoclonus** (quick, involuntary jerks)
- **Cortical sensory loss** (difficulty with recognising objects or sensations, not explained by primary sensory deficits)
- **Apraxia**, especially ideomotor apraxia (difficulty with purposeful movements despite normal strength and understanding)
- **Alien limb phenomenon**, where a limb moves involuntarily, often outside of the patient's awareness or control

Tremor, if present, is usually mild or absent. Gait disturbances and postural instability may develop as the disease progresses.

Cognitive and Neuropsychological Features in Corticobasal Degeneration

Cognitive changes in CBD may be as disabling as the motor deficits, and frequently present early:

- **Executive dysfunction**: Impaired planning, organisation, flexible thinking, and inhibition are common.
- **Language difficulties**: Non-fluent/agrammatic primary progressive aphasia may develop, with hesitant, effortful, and grammatically incorrect speech. Naming difficulties and word-finding pauses are also typical.
- **Visuospatial impairment**: Difficulties with spatial orientation, copying, and constructional praxis, sometimes related to parietal lobe involvement.
- **Behavioural symptoms**: Apathy, irritability, impulsivity, and, more rarely, personality change.
- **Memory**: Relative preservation of straightforward memory, but difficulties may arise from executive rather than storage deficits. Rapid forgetting or prominent learning difficulties are less characteristic than in Alzheimer's disease.

Alien limb phenomenon and pronounced apraxia can complicate both daily functioning and neuropsychological assessment, particularly in tasks that require complex hand movements or coordinated responses.

Differential Diagnosis in Corticobasal Degeneration

CBD can be mistaken for several other neurodegenerative disorders, especially early on:

- **Progressive Supranuclear Palsy (PSP)**: Can present with falls, vertical gaze palsy, and frontal/executive dysfunction; tends to have more symmetrical symptoms and less prominent limb apraxia or alien limb phenomena than CBD.
- **Parkinson's Disease**: Features slower progression, a greater response to levodopa, and more symmetrical symptoms.
- **Multiple System Atrophy (MSA)**: Distinguished by autonomic and cerebellar signs.
- **Alzheimer's Disease**: Early, prominent amnesia and less marked motor symptoms; some overlap in language and visuospatial deficits.

- **Frontotemporal Dementia (FTD)**: Can have overlapping features, particularly if parietal or frontal regions are involved.
- **Corticobasal Syndrome (CBS)**: A clinical syndrome that may be caused by CBD, but also by other underlying pathologies (like Alzheimer's or PSP), highlighting the complexity of diagnosis.

Careful assessment of the evolution and prominence of motor, cognitive, and behavioural features is crucial in distinguishing CBD from these mimics.

Neuropathology and Brain Imaging in Corticobasal Degeneration

CBD is defined pathologically by **asymmetric neuronal loss and tau-positive inclusions in the cortex** (especially frontoparietal) **and subcortical structures** (basal ganglia, substantia nigra). **Ballooned neurons** ("achromatic neurons") are a classic histological feature.

Brain imaging often shows **asymmetric cortical atrophy**, notably in the posterior frontal and parietal lobes, and **atrophy in the contralateral basal ganglia**. However, **overlap with other pathologies limits the diagnostic certainty from imaging alone**.

Embedded media: https://radiopaedia.org/cases/185756/play?lang=gb

PET, CT and MRI imaging of a real corticobasal degeneration case in a 70-year old with "diagnosis almost certain" © Radiopaedia

The Neuropsychologist's Role in Corticobasal Degeneration

Clinical Neuropsychologists have a central function in the management of CBD:

- **Assessment**: Documenting cognitive and behavioural symptoms, differentiating between storage and executive-mediated memory issues, clarifying features such as apraxia, alien limb, and language disturbance.
- **Diagnosis**: Assisting the multidisciplinary team to distinguish CBD from other neurological conditions through detailed cognitive profiling and collateral history.
- **Management**: Supporting individuals and families in managing cognitive, language, and behavioural problems, and helping set appropriate expectations for disease progression.

- **Adaptation and Rehabilitation**: Suggesting compensatory strategies and environmental adaptations to maintain independence and quality of life for as long as possible.

Summary

Corticobasal Degeneration is a rare neurodegenerative illness distinguished by an unusual combination of asymmetric movement disorder, apraxia, alien limb phenomenon, and progressive cognitive and language dysfunction. Neuropsychologists play a crucial role in early detection, differential diagnosis, and ongoing support for people affected and their families. Always consider CBD in the differential when a patient presents with a combination of asymmetry, apraxia, executive or language dysfunction, and unusual limb phenomena.