

# Corticobasal Degeneration

## ISSUE BRIEF ON CORTICOBASAL DEGENERATION

### Introduction

Briefings such as this one are prepared in response to petitions to add new conditions to the list of qualifying conditions for the Minnesota medical cannabis program. The intention of these briefings is to present to the Commissioner of Health, to members of the Medical Cannabis Review Panel, and to interested members of the public scientific studies of cannabis products as therapy for the petitioned condition. Brief information on the condition and its current treatment is provided to help give context to the studies. The primary focus is on clinical trials and observational studies, but for many conditions there are few of these. A selection of articles on pre-clinical studies (typically laboratory and animal model studies) will be included, especially if there are few clinical trials or observational studies. Though interpretation of surveys is usually difficult because it is unclear whether responders represent the population of interest and because of unknown validity of responses, when published in peer-reviewed journals surveys will be included for completeness. When found, published recommendations or opinions of national organizations medical organizations will be included.

Searches for published clinical trials and observational studies are performed using the National Library of Medicine's MEDLINE database using key words appropriate for the petitioned condition. Articles that appeared to be results of clinical trials, observational studies, or review articles of such studies, were accessed for examination. References in the articles were studied to identify additional articles that were not found on the initial search. This continued in an iterative fashion until no additional relevant articles were found. Finally, the federal government-maintained web site of clinical trials, [clinicaltrials.gov](http://clinicaltrials.gov), was searched to learn about trials currently under way or under development and to check whether additional articles on completed trials could be found.

### Definition

Corticobasal degeneration (CBD) is a rare, progressive neurodegenerative condition due to pathological accumulation in brain neurons of tau protein. Patients with a combination of symptoms suggestive of the disorder are often referred to as having 'corticobasal syndrome' (CBS), while CBD is used strictly to describe cases verified by pathology studies after death. Clinical diagnosis of CBD can be difficult as symptoms are variable and often resemble those of other types of neurodegenerative disorders (Armstrong 2016).

Patients typically develop symptoms in their 6<sup>th</sup> or 7<sup>th</sup> decades with a mean age of onset of 64 years and mean survival of 6 to 7 years (Lamb 2016).

The most common presenting symptom is limb clumsiness affecting one side of the body, initially with or without accompanying rigidity or tremor. Subsequently, the disease spreads to affect gait and there is slow progression to influence the arm and leg on the same side where symptoms first appeared. Eventually, major clinical features include apraxia

(difficulty carrying out intended movements) and dementia, parkinsonism (tremor, slow movement, and rigidity), palsy (weakness or paralysis), and myoclonus (spasmodic contractions of muscle groups) (Armstrong 2016).

## Prevalence

Prevalence of CBD is estimated at 4.9 – 7.3 per 100,000 (Mahapatra 2004).

## Current Therapies

Despite many efforts, disease-modifying treatment is not yet available for CBD. Active research is ongoing to find treatments for CBD and related neuropathological disorders (Marsili 2016).

In the absence of disease-modifying therapies for CBD, management is based on relieving symptoms and assisting patients with their activities of daily living. Advanced care planning and non-pharmacological supportive therapies are very important. Non-pharmacological supportive therapies include diet consultation (due to frequency of swallowing difficulty and poor appetite), physical therapy to safely maintain strength and balance, and speech and occupational therapy. Most patients will be trialed on L-DOPA and amantadine, although there is only limited evidence that some patients may experience modest improvement in Parkinsonism with these drugs. A wide variety of other drugs are used to treat symptoms common in CBD. Examples include sertraline and citalopram (for depression), clomipramine and trazodone (for depression and behavioral symptoms), and diazepam (for dystonia and myoclonus) (Lamb 2016).

## Pre-Clinical Research

No preclinical studies of cannabis or cannabinoids relevant to CBD or related neurodegenerative disorders were found.

## Clinical Trials

No clinical trials of cannabis or cannabinoids relevant to CBD or related neurodegenerative disorders were found.

## Observational Studies

No observational studies of cannabis or cannabinoids relevant to CBD or related neurodegenerative disorders were found.

## National Medical Organization Recommendations

No guidance documents or recommendations from national medical organizations for the therapeutic use of cannabis or cannabinoids in the management of endocannabinoid deficiency were found.

## References

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