

# Cortiocbasal Degeneration 101

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# Disclosures

- I have nothing to declare
- I will try not to use too many medical terminologies

 **Welcome to Canada**  
**Your Customs Declaration Card – Completion Guide**

This guide gives you information about rules and exemptions that may apply to you and the goods you are bringing into Canada. The following information corresponds to the sections on the attached Customs Declaration Card.

**Part A – All travellers**

All travellers must be identified on a Customs Declaration Card. You may list up to five family members living at the **same address** on one declaration card. Please ensure that each traveller's answers are included on the card. **Each traveller is responsible for his or her own declaration.**

**You must declare all goods that you are bringing into Canada, whether or not they are part of your exemption.** A customs officer may examine your goods. It is important to note that the value of goods that do not qualify for a personal exemption may be subject to duties, taxes, and/or fees.

**Customs officers may seize any goods you do not declare or any goods you falsely declare. You may also face prosecution.**

Goods like the following ones may be **restricted, controlled, or prohibited** in Canada, or you may need a **special admission permit**. Failure to declare all of these items could result in fines, arrest, and prosecution:


- Firearms or other weapons (e.g., mace, switchblades, fireworks);
- Drugs;
- Endangered species (live specimens or products made from wild plants or animals, e.g., ivory, corals, reptile skin products); and
- Cultural property (e.g., antiques, cultural objects).

Under Canadian law, as administered by the **Canadian Food Inspection Agency**, you have to declare all animals and plants and their products. Failure to do so could result in fines and prosecution.

There are specific personal exemption limits for importing alcohol and tobacco into Canada. Determine your limit from the following table.

Minimum age	Province or territory	Alcohol and tobacco
18 years	Alberta, Manitoba, Quebec	1.5 L of wine or 1.14 L of liquor or 24 x 355 mL (12 oz.) bottles/cans of beer or ale (8.5 L)
19 years	All others	
18 years	Alberta, Manitoba, Quebec, Saskatchewan, Yukon Territory, Northwest Territories, Nunavut	200 cigarettes, 200 tobacco sticks, 50 cigars or 50 cigarillos, and 200 grams manufactured tobacco
19 years	All others	

During your travels, you may have been exposed to infectious diseases not common in Canada. While the risks posed by most of these illnesses are low, some, such as malaria, may be life threatening. If you develop a fever or flu-like illness within three months of travel to a tropical country, **Health Canada** advises you to get immediate medical attention, inform your doctor of your travel history, and ask for a blood test to rule out malaria.

E311 (00) (Français au verso) 

E311 (00) (Français au verso) 



# Outline

1. What is corticobasal degeneration (CBD)
2. Management

# THE STORIES OF 3 PEOPLE

# Case

## 72 year-old right-handed widowed man

Timeline	Story
5 years ago	Presented with executive dysfunction (planning and multitasking difficulties) and some short-term memory difficulties. Normal neurological examination.
2-4 years	He was stable over the subsequent years. Moved to a retirement home.
1-2 years	Started to have difficulties with walking – had difficulties standing, complained of weakness on 1-side and slowness. Neurological examination revealed weakness on the left side in a “stroke-like” pattern and stiffness/rigidity. He was slow with his thinking and movements – not due to the weakness.
	Passed away from a pneumonia

# Case

## 65 year-old right-handed married man

Timeline	Story
4 years ago	Developed word-finding difficulties; appeared to “stutter” when tried to say a word
3 years ago	Developed slowness and stiffness – mainly on the right side, shuffled when he walks
2.5 years ago	Developed weakness on the right-side of the body in a “stroke-like” pattern over time
1 year	Unable to ambulate; is dependent on others for his day-to-day care; marked rigidity and spasticity in the arms and legs

# Case

## 62 year-old left-handed woman

Timeline	Story
4 years ago	Developed visuospatial difficulties – could not find things in front of her, having problems navigating/finding her way in familiar places
3 years ago	Found it difficult to read as the letters appear to jump around the page; when reaching for items, could not “see” them and would miss
1 year ago	Started to “ignore” the left side of her body; started to use her right hand more. Neurological examination revealed visuospatial difficulties, mild rigidity and asymmetric parkinsonism, and mild “stroke-like” pattern of weakness on her left side

# What Do These Cases Have in Common?

Their final diagnosis:

**Corticobasal Degeneration**



# But They All Seem Different...

- All cases started differently
- All cases had different symptoms
- But all cases had some similar features
- Yet, they are all caused by corticobasal degeneration (CDB)....so what it is?

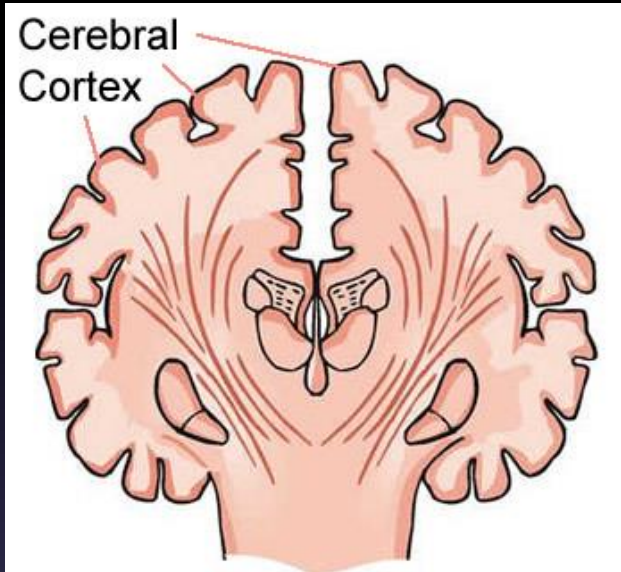
# What is Corticobasal Degeneration (CBD)?

**Cortico**

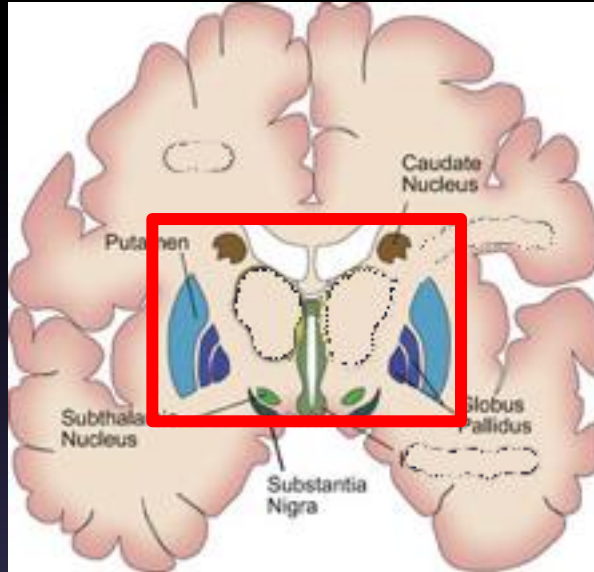


**Basal**

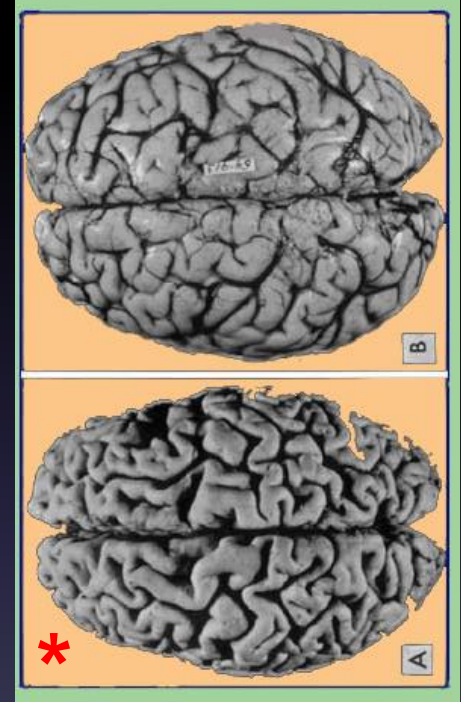
**Degeneration**



Involvement of the **cortex** (thinking part of the brain; dysfunction causes cognitive impairment & dementia)



Involvement of the **basal** ganglia (movement of the body; dysfunction causes lack of movement (Parkinson's disease) or too much movement (Huntington's disease))



Atrophy\* or shrinkage of the brain

# Epidemiology of CBD

- Incidence (new cases)
  - 1/100 000\* per year
- Age Distribution
  - Range usually 40 – 80 years old
- Gender distribution
  - No clear difference between women and men – both are equally affected
- Environmental Risk Factors
  - None known
- Duration of disease/  
symptom progression
  - 3 to 15 years
- Genetics
  - Some have mutation in *MAPT* gene (chr 17) or *PGRN* (chr 17)

# Classic Presentation of CBD

**Asymmetric rigidity**  
(parkinsonism)



**Apraxia**  
(inability to perform the learned motor programming of tasks)



**Alien limb**  
(a limb – arm or leg – is perceived as not belonging to the same person)



**Myoclonus**  
(spontaneous jerking of a muscle)

# Classic Presentation of CBD

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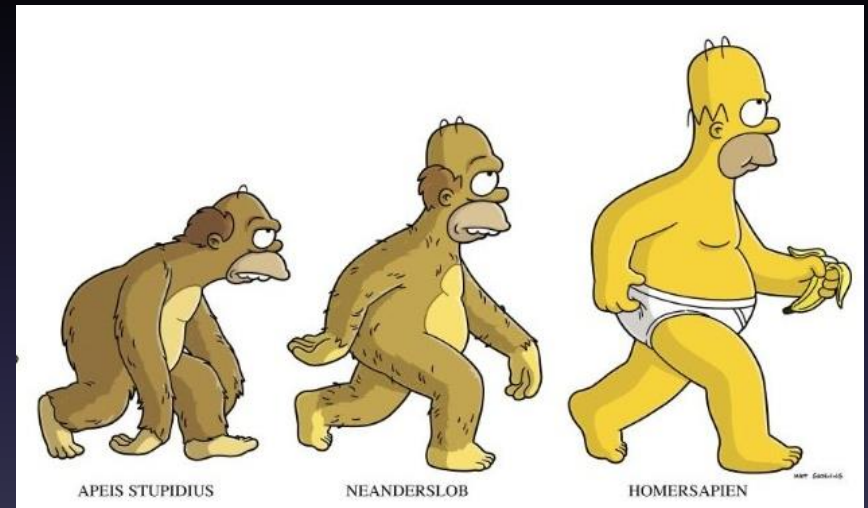


**Myoclonus**  
(spontaneous jerking of a muscle)

**Unfortunately, this is not the most common presentation and thus making a diagnosis can be challenging**

# Difficulties to Diagnose CBD

- First described in 1967 in 3 persons with the disorder
- Over the next 50 years, there has been an evolving knowledge of this disorder



# Difficulties to Diagnose CBD

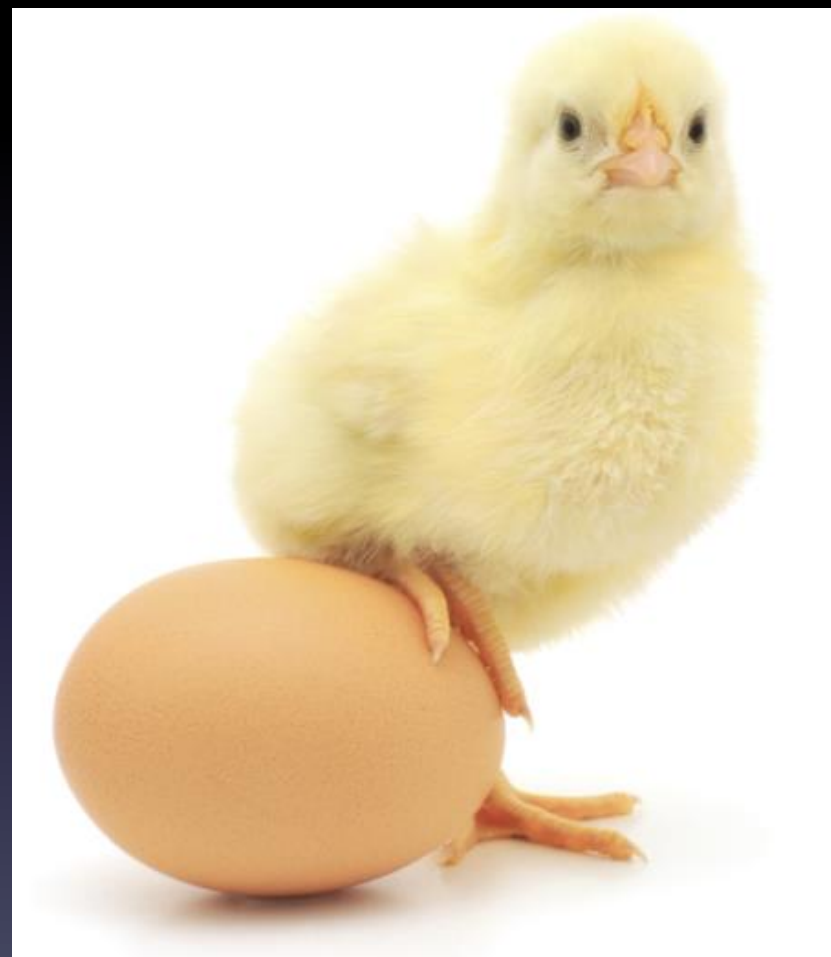
- Which came first?

Parkinsonism?

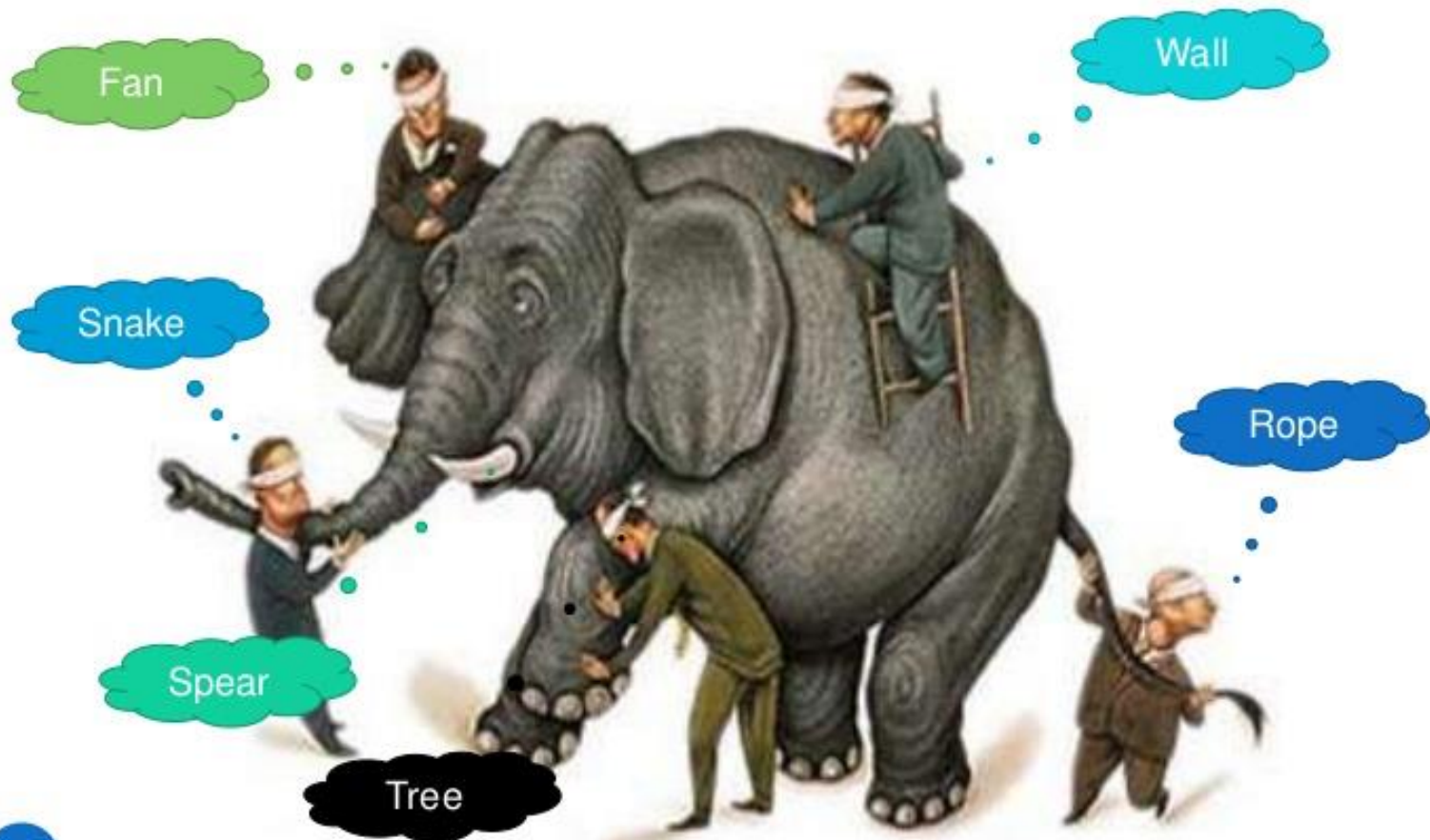


Cognitive impairment?

- Initially, it was thought that parkinsonism came first and cognitive disorders was not common



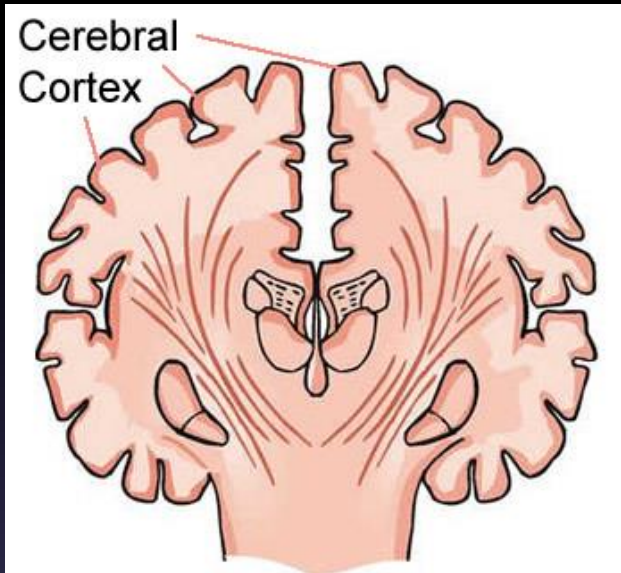
The elephant metaphor:  
poem by John Godfrey Saxe (1816-18-87)



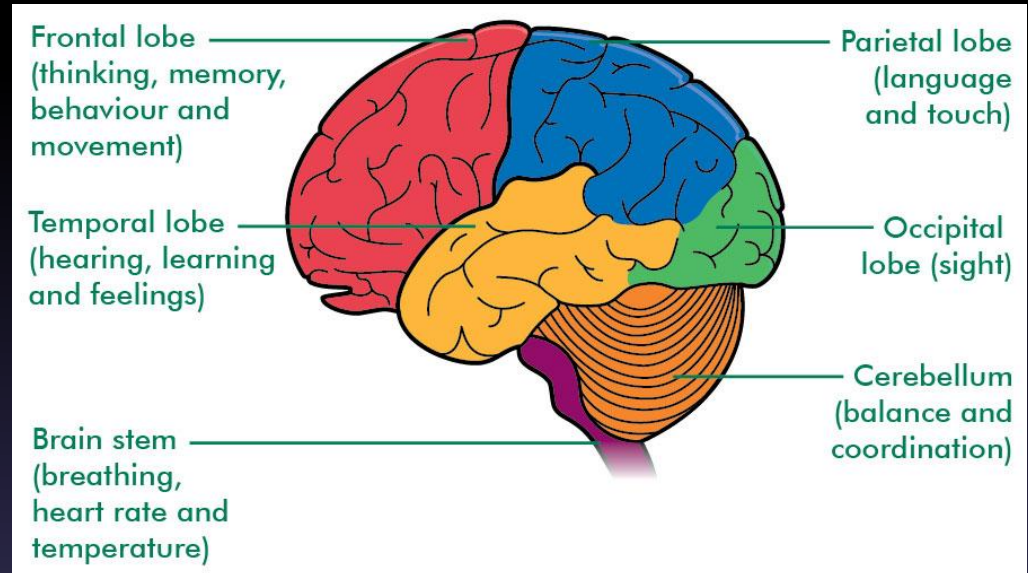


# Difficulties to Diagnose CBD

## Cortico



Involvement of the **cortex** (thinking part of the brain; dysfunction causes cognitive impairment & dementia)

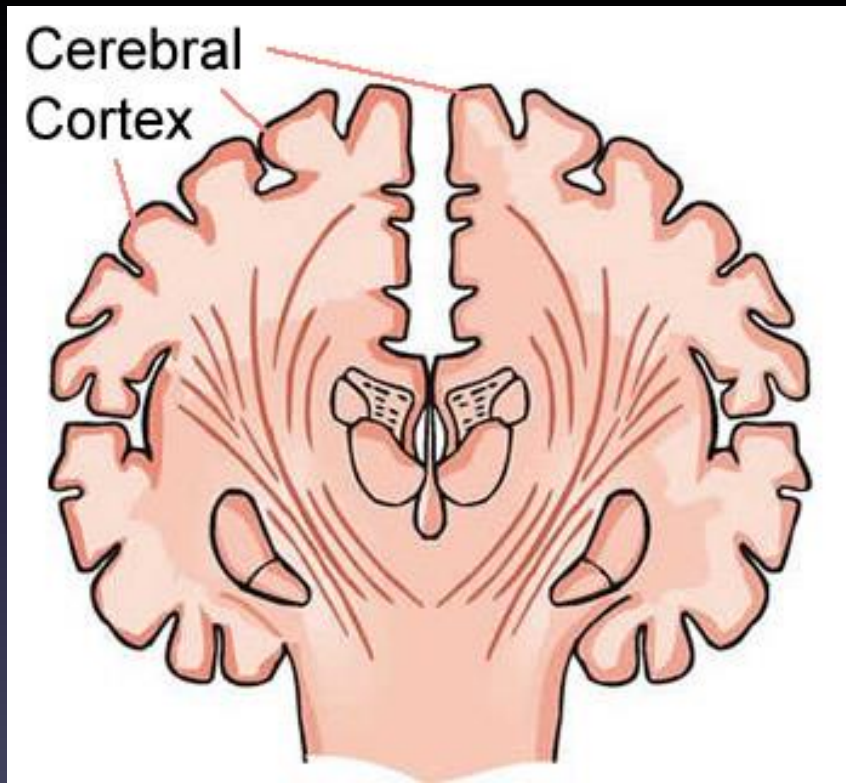


Since the brain has many functions (above), CBD can present/start with difficulty in a single cognitive domain and evolve over time.

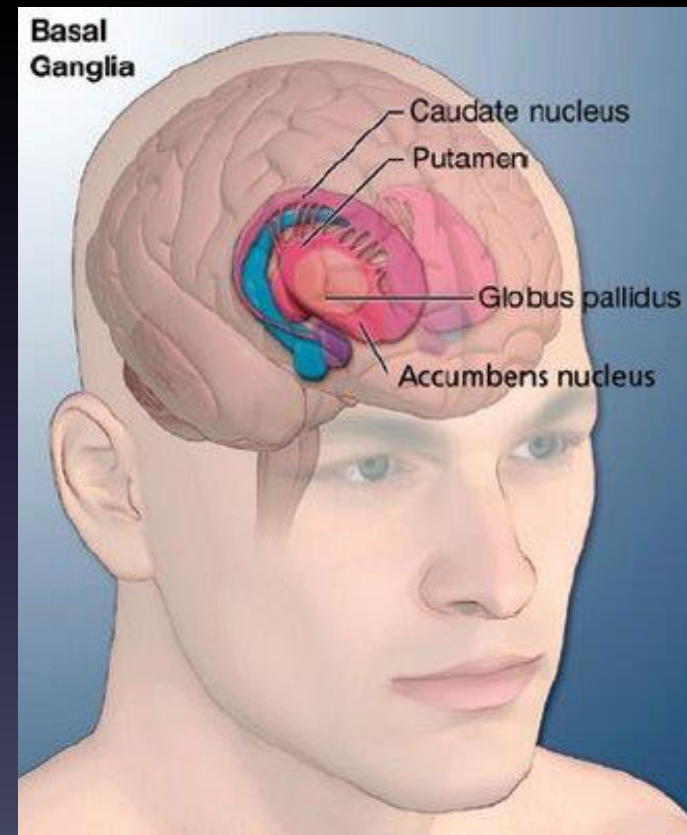
# How to Diagnose CBD

*Back to Basics*

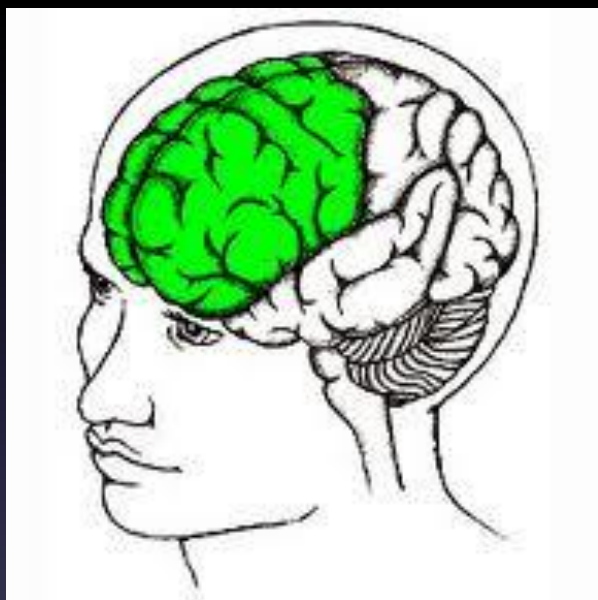
## Cognitive/Thinking Features



## Motor Features



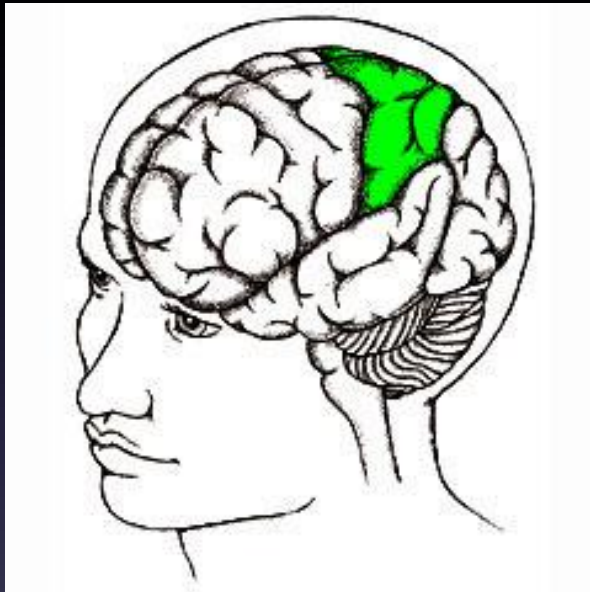
# Cognitive Symptoms of CBD



**Frontal Lobe**

- **Executive dysfunction**
  - Planning and multitasking difficulties
- **Behavioural changes**
  - Disinhibition, hypersexual, apathetic, antisocial

# Cognitive Symptoms of CBD



**Parietal Lobe**

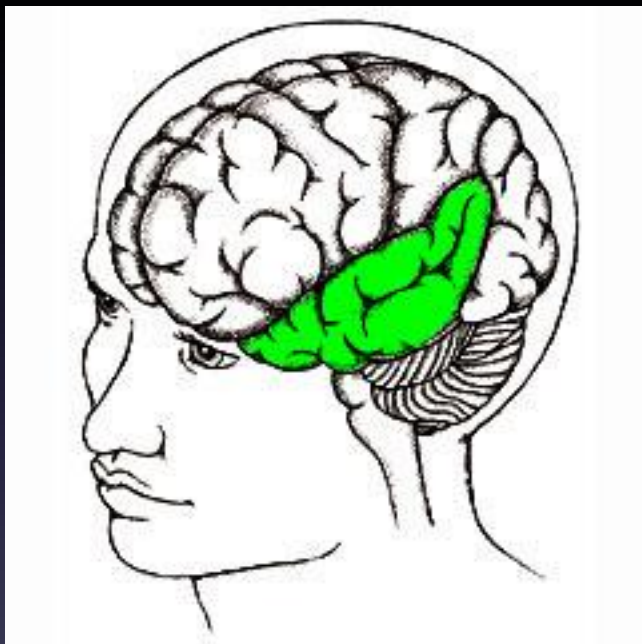
- **Apraxia**
  - “not knowing how to do it”
- **Cortical sensory loss**
  - Inability to recognize objects based on touch eventhough sensation is normal (stereognosis)
  - Inability to recognize written figures on hand (graphesthesia)
  - Inability to recognize one side of the body (extinction)

# Cognitive Symptoms of CBD



- Alien limb phenomenon
  - “limb has a mind of it’s own”
  - “this [hand] does not belong to me”

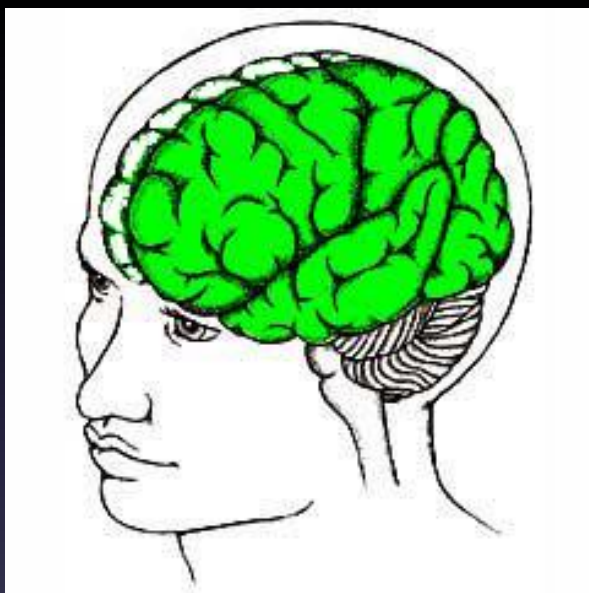
# Cognitive Symptoms of CBD



Temporal Lobe

- Short-term memory loss (similar to Alzheimer's disease)

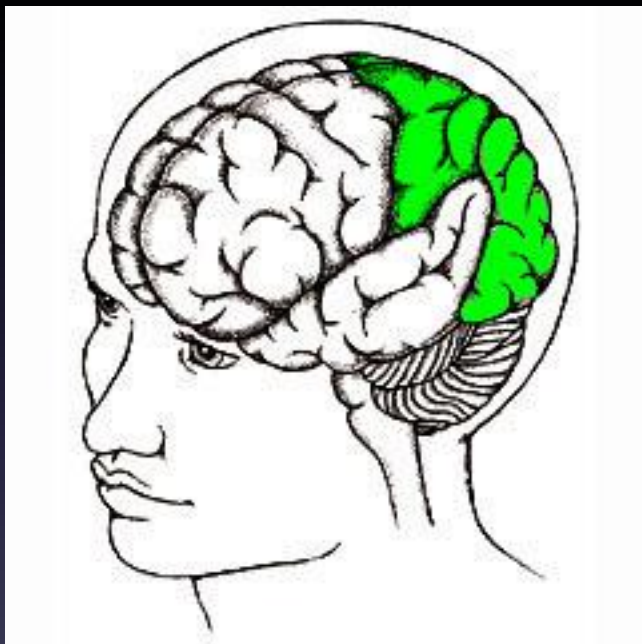
# Cognitive Symptoms of CBD



**Left Hemisphere**

- Language –  
progressive aphasia  
(non-fluent subtype)
  - Word-finding difficulties  
(anomia)
  - Pronunciation difficulties  
(oral apraxia)
  - Loss of grammar

# Cognitive Symptoms of CBD

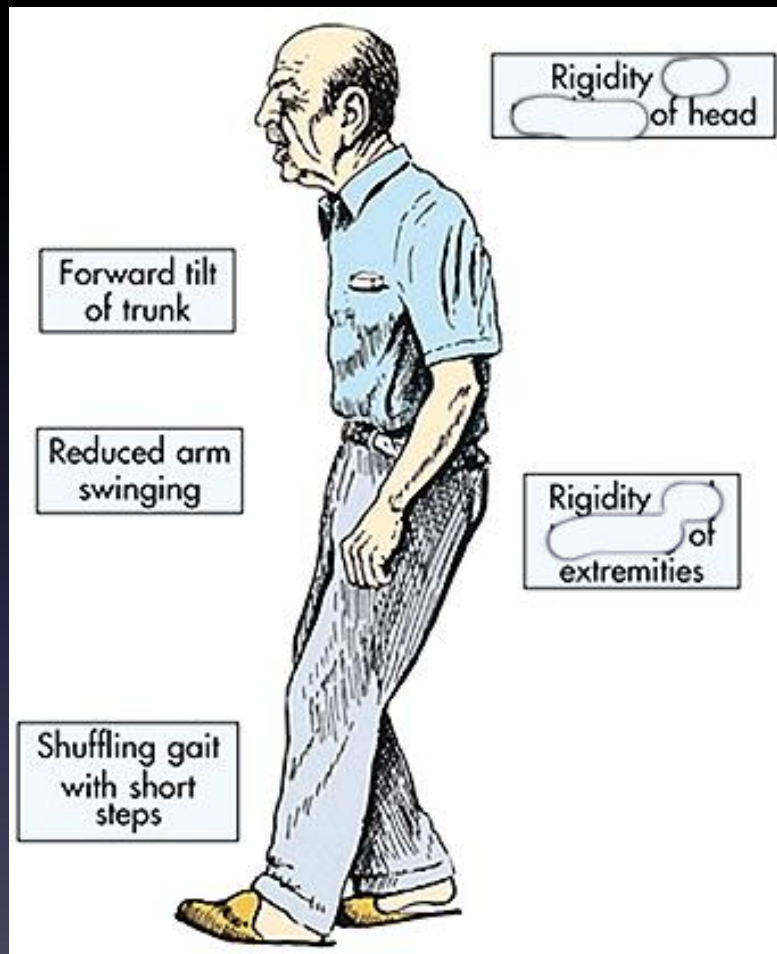


**Parietal & Occipital Lobes**

- **Visuospatial impairment**
  - Presentation similar to PCA (simultanagnosia, optic ataxia, ocular apraxia)



# Motor Features of CBD



- **Parkinsonism**

- *Asymmetric* – one side is more affected than the other
- Tremor is *not* commonly seen
- Most persons *do not* respond to levodopa, a Parkinson's medication)
  - Small minority there is a transient response

- **Bradykinesia** (overall slowness)

# Clarification:

## Parkinsonism is NOT Parkinson's Disease

SYMPTOMS

### Parkinsonism

Describes a collection of symptoms that include rigidity (stiffness), bradykinesia (slow movement), balance/posture changes, tremor

CAUSES

Parkinson's disease

Medications  
(antipsychotics,  
antiemetics)

Strokes

Corticobasal  
Degeneration

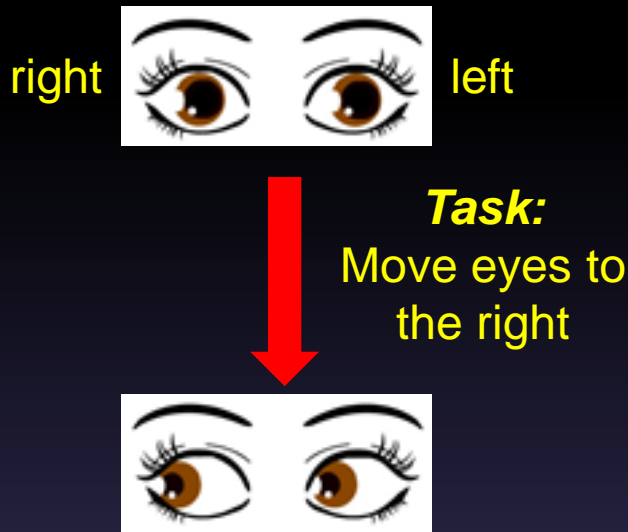
Progressive  
Supranuclear  
Palsy

Multiple  
System  
Atrophy

# CBD Compared to Other Parkinsonian Disorders

	Parkinson's Disease	Dementia with Lewy Bodies	Multiple System Atrophy	Progressive Supranuclear Palsy	Corticobasal Degeneration
<i>Tremor</i>	+	+/-	-	-	-
<i>Rigidity</i>	<b>+ (limb)</b>	<b>+ (limb)</b>	<b>+ (limb)</b>	<b>+ (axial&gt;&gt;limb)</b>	<b>+ (limb, very asymmetric)</b>
<i>Akinesia</i>	+	+	+	+	+
<i>Postural Instability</i>	+/-	+/-	++	+++	+/-
<i>Bradyphrenia</i>	+	+	+	+	+
<b>Response to Levodopa</b>	<b>+</b>	<b>+/-</b>	<b>-</b>	<b>-</b>	<b>-</b>
<b>Hallmark Features</b>	<ul style="list-style-type: none"> <li>• REM sleep behaviour disorder (RBD)</li> <li>• Autonomic disturbance (late)</li> <li>• Dysphagia (late)</li> <li>• Cognitive impairment usually occurs later</li> </ul>	<ul style="list-style-type: none"> <li>• <b>Dementia with spontaneous visual hallucinations and fluctuations</b></li> <li>• RBD</li> <li>• Autonomic disturbance (late)</li> <li>• Dysphagia (late)</li> </ul>	<ul style="list-style-type: none"> <li>• RBD</li> <li>• <b>Early autonomic dysfunction</b> (orthostatic hypotension, postprandial hypotension, anhidrosis with thermoregulatory disturbances, constipation, impotence, poor lacrimation and salivation)</li> <li>• <b>Urinary incontinence (early)</b></li> <li>• Cerebellar findings (ataxia, kinetic tremor) - MSA-C</li> <li>• <b>Nocturnal stridor</b></li> <li>• <b>NO DEMENTIA</b></li> </ul>	<ul style="list-style-type: none"> <li>• Marked postural instability cause <b>early falls</b></li> <li>• <b>Vertical ocular gaze paresis</b></li> <li>• Slow saccades</li> <li>• Applause sign (clapping after the patient has been instructed to clap 3 times)</li> </ul>	<ul style="list-style-type: none"> <li>• <b>Ideomotor apraxia</b></li> <li>• Alien limb</li> <li>• Marked asymmetry of signs</li> <li>• Cortical sensory loss</li> <li>• <b>Progressive aphasia</b> (non-fluent primary progressive aphasia presentation)</li> </ul>

# Motor Features of CBD



## **2 variables to measure:**

1. Time it takes to act on the task
2. Time it takes for the eye to move from midposition to the right

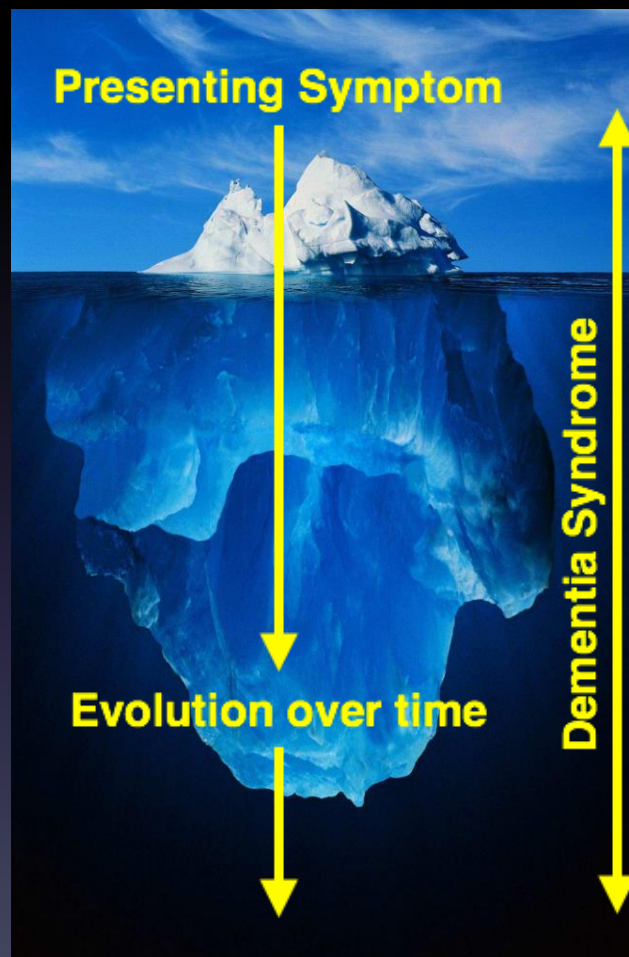
- **Eye movement changes**
  - (Figure to the left)
  - Slow to start an eye movement but the speed (velocity is normal)
- **Dysphagia** (difficulty swallowing)
  - Usually occurs as the disease progresses

# Motor Features of CBD

- **Myoclonus** (intermittent muscle jerks)
  - Stimulus sensitive
- **Dystonia** (abnormal co-contractions of muscle groups)
  - Usually affecting the arm
- **Upper motor neuron findings** (motor stroke symptoms except not sudden in onset)

# How To Determine If CBD

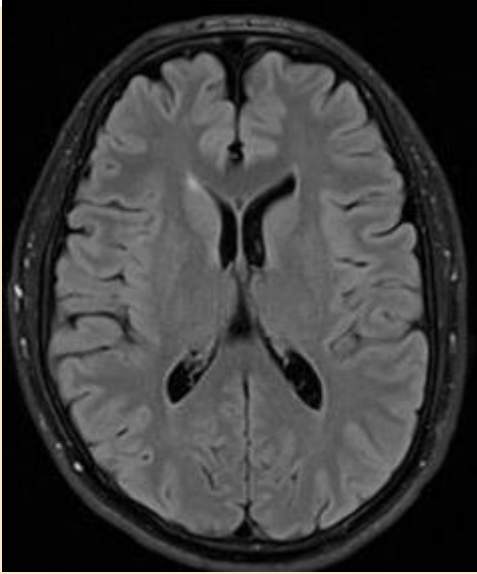
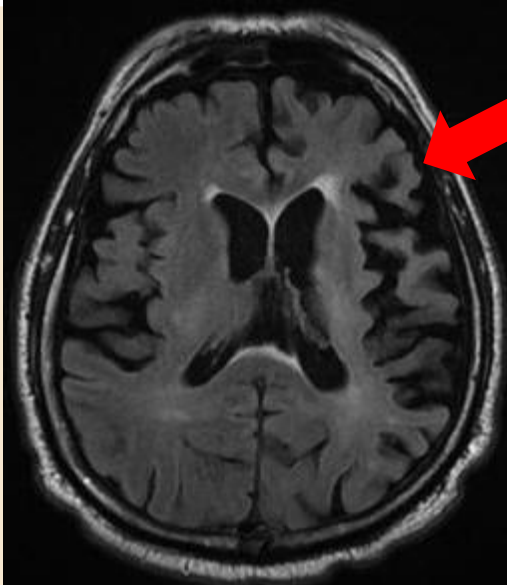
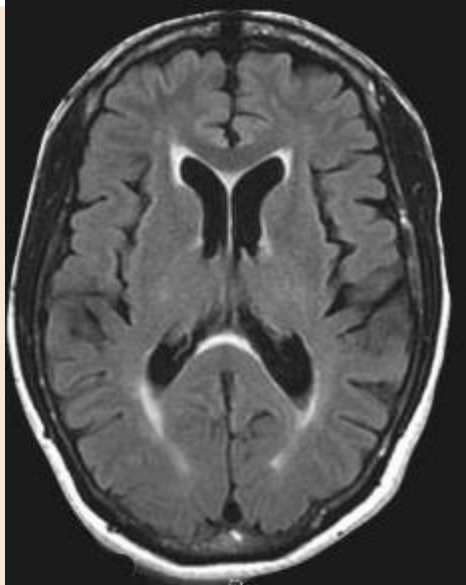
- Timing of symptoms
  - What started first, then second, then third etc.
- Follow-up and evolution over time



# CBD vs AD

	Corticobasal Degeneration	Alzheimer's Disease
Primary cognitive symptom	<ul style="list-style-type: none"><li>• Language (loss of words)</li><li>• Praxis</li></ul>	Short-term memory loss
Motor/physical symptoms	Parkinsonism (slowness, asymmetric)	None
Tremor	None	None

# Imaging Features of CBD

Normal	Corticobasal Degeneration	Alzheimer's Disease
		
	<p>Asymmetric atrophy (shrinkage) of 1 hemisphere</p>	<p>Usually hippocampal (memory structures) atrophy</p>



# Is There A Cure?



Unfortunately...no

# Treatment

## Cortical/Cognition

- Activity

## Basal/Motor

- Parkinson's medications (levodopa)
  - Titrate to high doses (1200 mg per day)
- Physiotherapy
- Occupational therapy

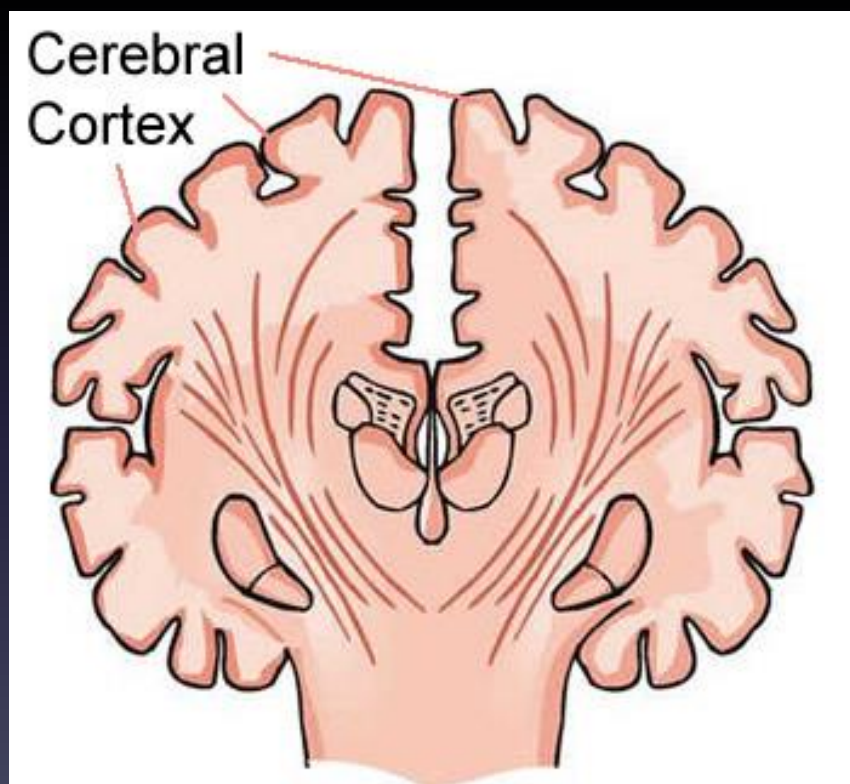
# SUMMARY

# Epidemiology of CBD

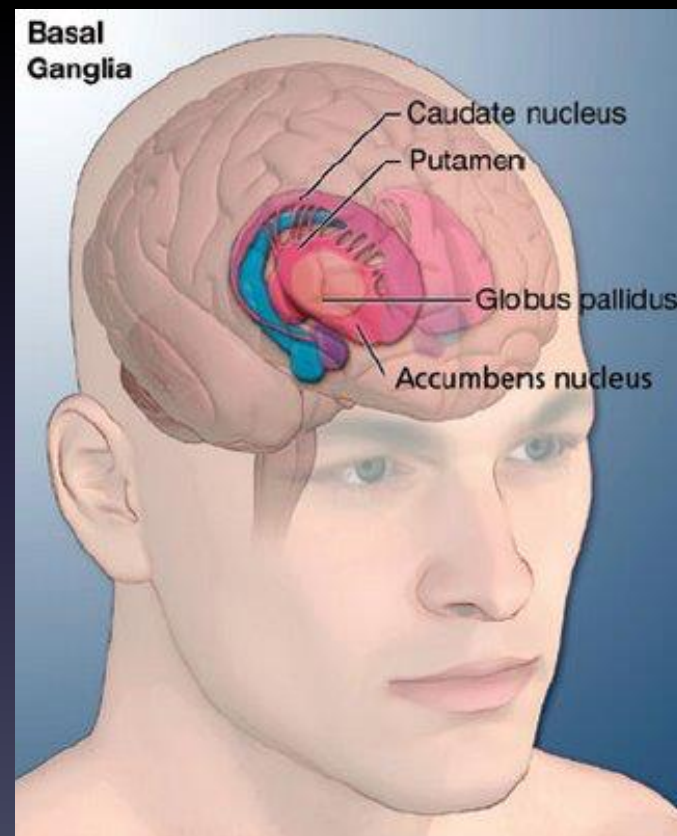
- Incidence (new cases)
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- Age Distribution
  - Range usually 40 – 80 years old
- Gender distribution
  - No clear difference between women and men – both are equally affected
- Environmental Risk Factors
  - None known
- Duration of disease/  
symptom progression
  - 3 to 15 years

# Clinical Features of CBD

## Cognitive/Thinking Features



## Motor Features



# Cortical Symptoms of CBD

- The “A’s”
  - **Apraxia**
    - “not knowing how to do it”
  - **Aphasia (non-fluent subtype)**
    - Word-finding difficulties (anomia)
    - Pronunciation difficulties (oral apraxia)
    - Loss of grammar
  - **Alien limb phenomenon**
    - “limb has a mind of it’s own”
    - “this [hand] does not belong to me”
- **Visuospatial impairment**
  - Presentation similar to PCA
- **Cortical sensory loss**
  - Inability to recognize objects based on touch even though sensation is normal
- **Executive dysfunction**
  - Planning and multitasking difficulties
- **Behavioural changes**
  - Disinhibition, hypersexual, apathetic, antisocial

# Motor Features of CBD

- **Parkinsonism**
  - Asymmetric – one side is more affected than the other
  - Tremor is NOT commonly seen
  - Most persons do not respond to levodopa, a Parkinson's medication)
    - Small minority there is a transient response
- **Bradykinesia** (overall slowness)
- **Eye movement changes**
  - Slow to start an eye movement but the speed (velocity is normal)
- **Myoclonus** (intermittent muscle jerks)
  - Stimulus sensitive
- **Dystonia** (abnormal contractions of muscle groups)
  - Usually affecting the arm
- **Gait changes**
  - Shuffling gait with reduced arms swing, as seen in Parkinson's disease
- **Dysphagia** (difficulty swallowing)
- **Upper motor neuron findings** (motor stroke symptoms except not sudden in onset)

# Treatment

## Cortical/Cognition

- Activity

## Basal/Motor

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# Thank You

