Cognition and mood in PSP and CBS

Dr Susan O'Connell

Consultant Clinical Psychologist

INS

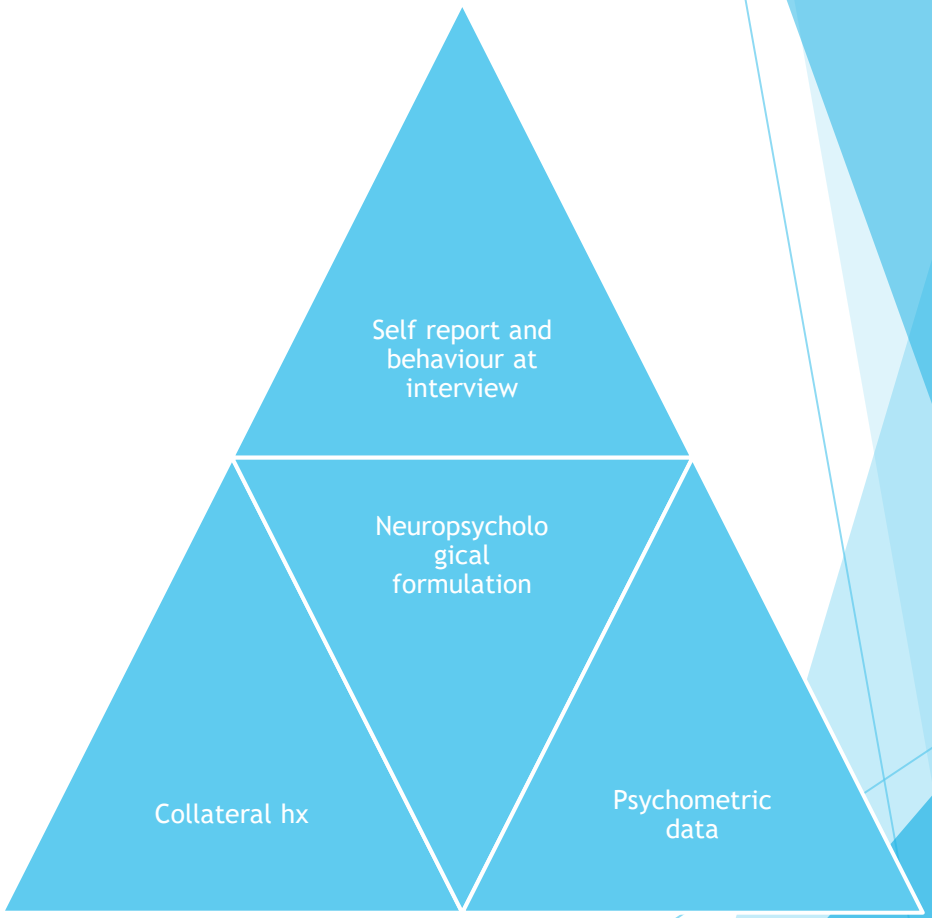
NHS Greater Glasgow & Clyde

Over view

- Why assess cognition?
- Brief orientation to the neuroanatomy of PSP and CBS
- Different cognitive symptoms of CBS and PSP
- Overlap of condition
- Rehab strategies
- Assessment
- Mood
- Management
- Signposting

Neuropsychology

How a known or yet undiagnosed neurological condition impacts on someone's cognition, emotions and behaviours

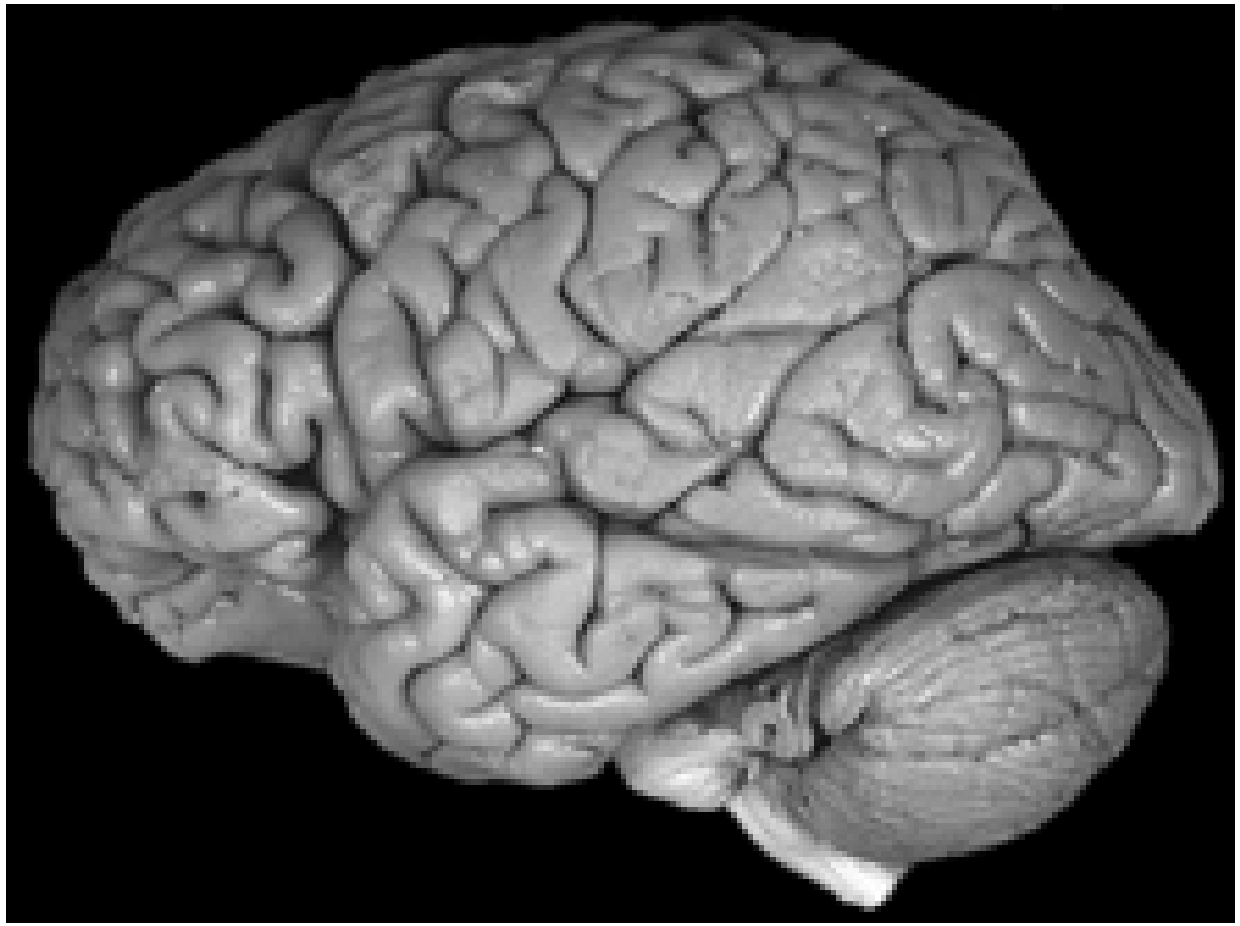


Impact of cognitive changes

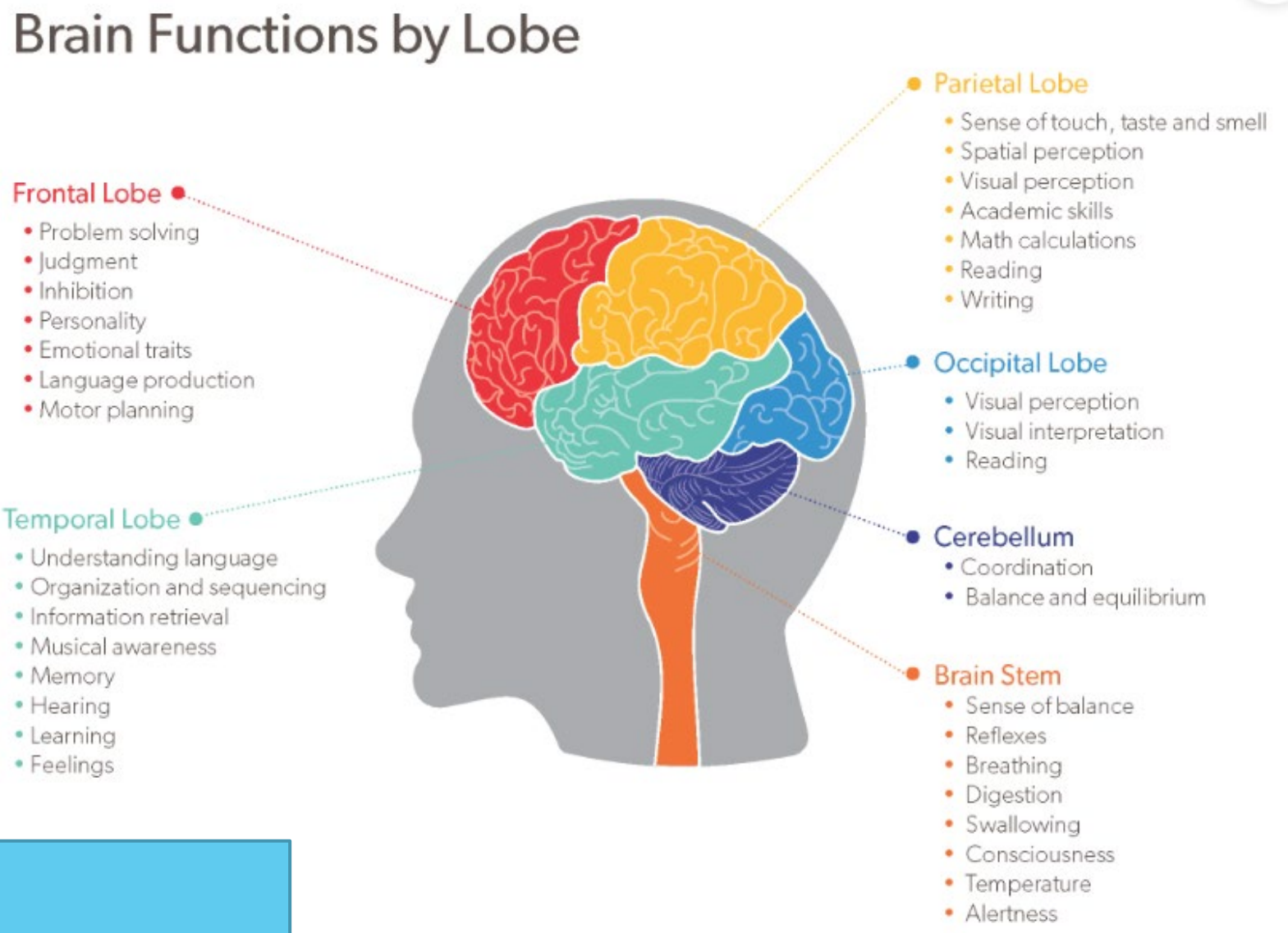
These significant changes impact on:

- Managing affairs and finances
- Independence and ADLs
- Planning for the future
- Difficulties making decisions (issues of capacity).
- Difficulties within family systems and relationships.
- Difficulties within care systems.

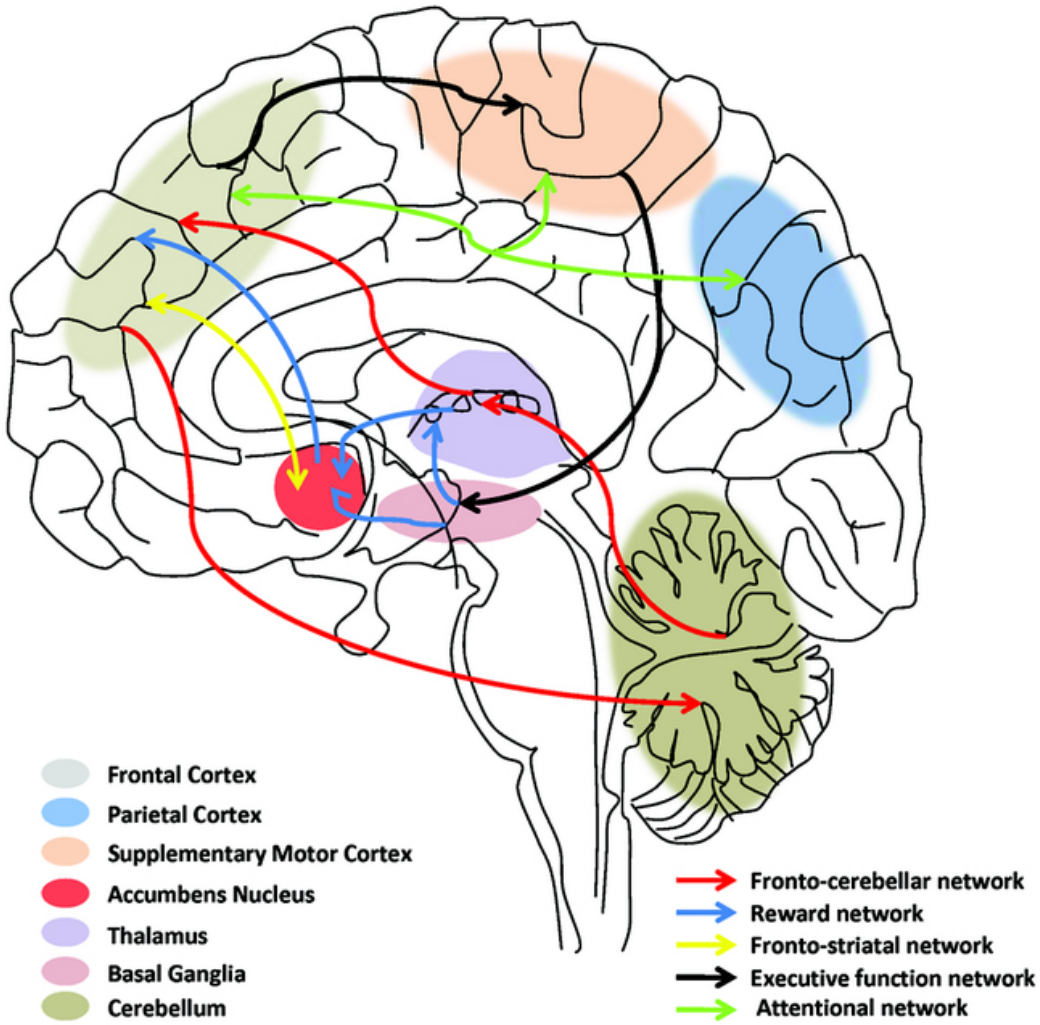
Cerebral function ***and*** connectivity



Cortical Regions



Cognition – subcortical and WMT



- Connectivity is important to remember
- Note how many project to frontal lobe which holds responsibility to executive functioning

Common subcortical cognitive changes

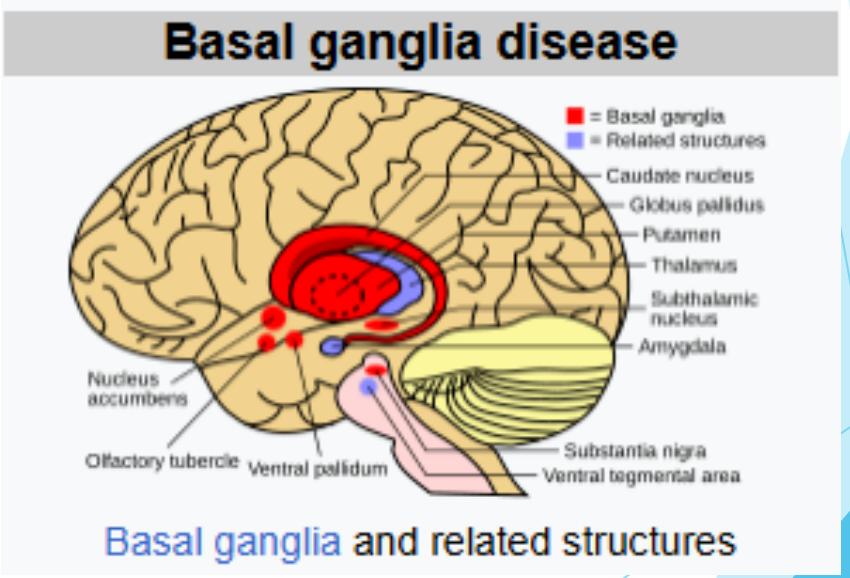
Processing speed

Attention

Basal ganglion circuits

There are 5 circuits that functionally and structurally connect the BG to cortex and thalamus

1. Motor
2. Occulomotor
3. Dorso-lateral
4. Orbito frontal
5. Medial frontal



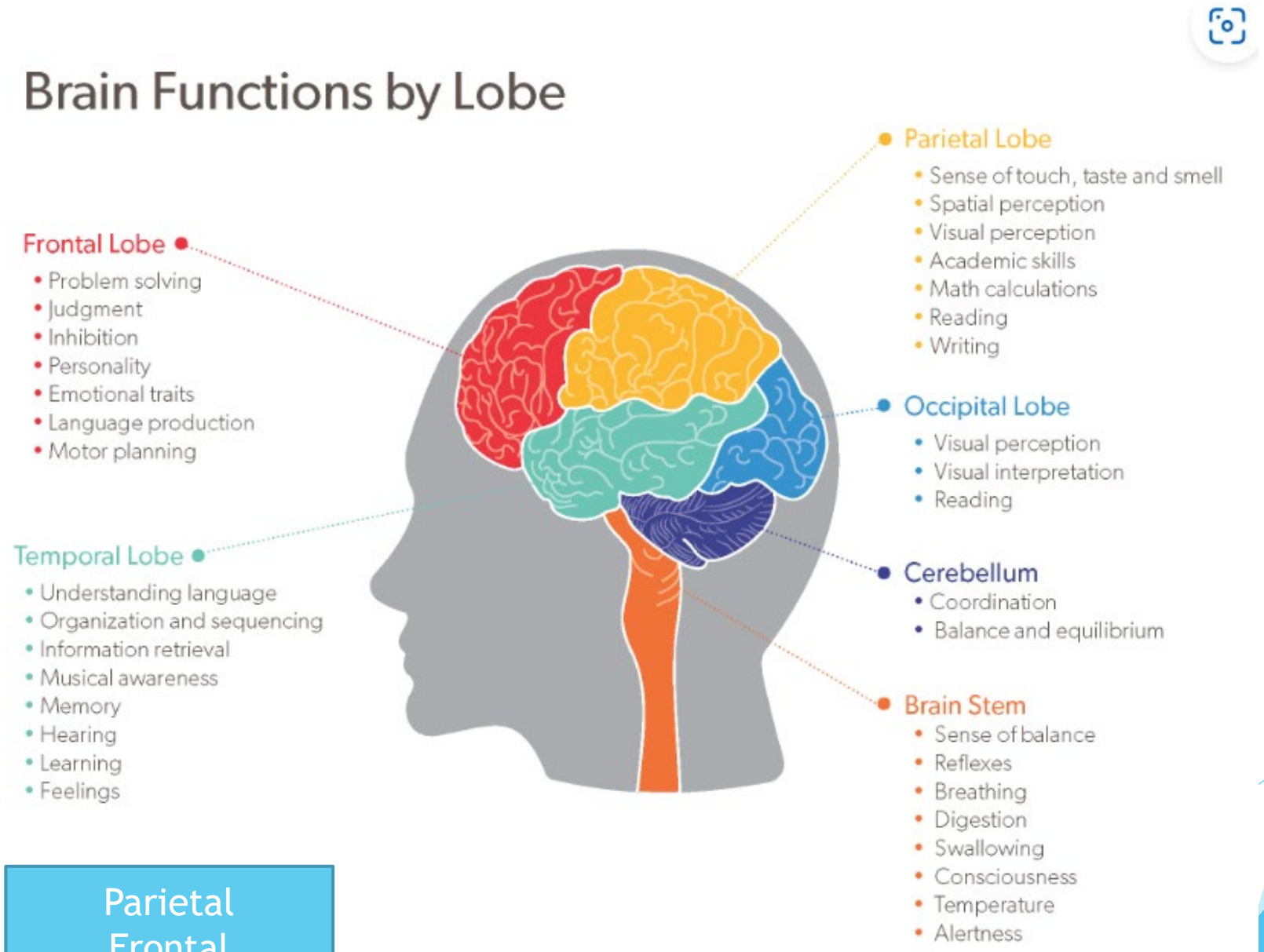
Cortical Basal Syndrome (CBS)

Cortical Basal Syndrome

- Accumulation of Tau protein greater in cortical sensory regions and putamen
- 4r Tauopathy
- MRI/SPECT asymmetrical findings in posterior parietal and frontal regions
- Asymmetrical onset of motor symptoms – parkinsonian in nature
- Severe ideomotor apraxia
- Alien limb in 60% (due to change in non dominant parietal lobe)
- Can have bulbar symptoms but later than PSP. If early bulbar symptoms then poorer prognosis
- Motor and cognitive symptoms can occur in equal measure even in early stages
- Left side symptoms at onset linked with greater cognitive impairment

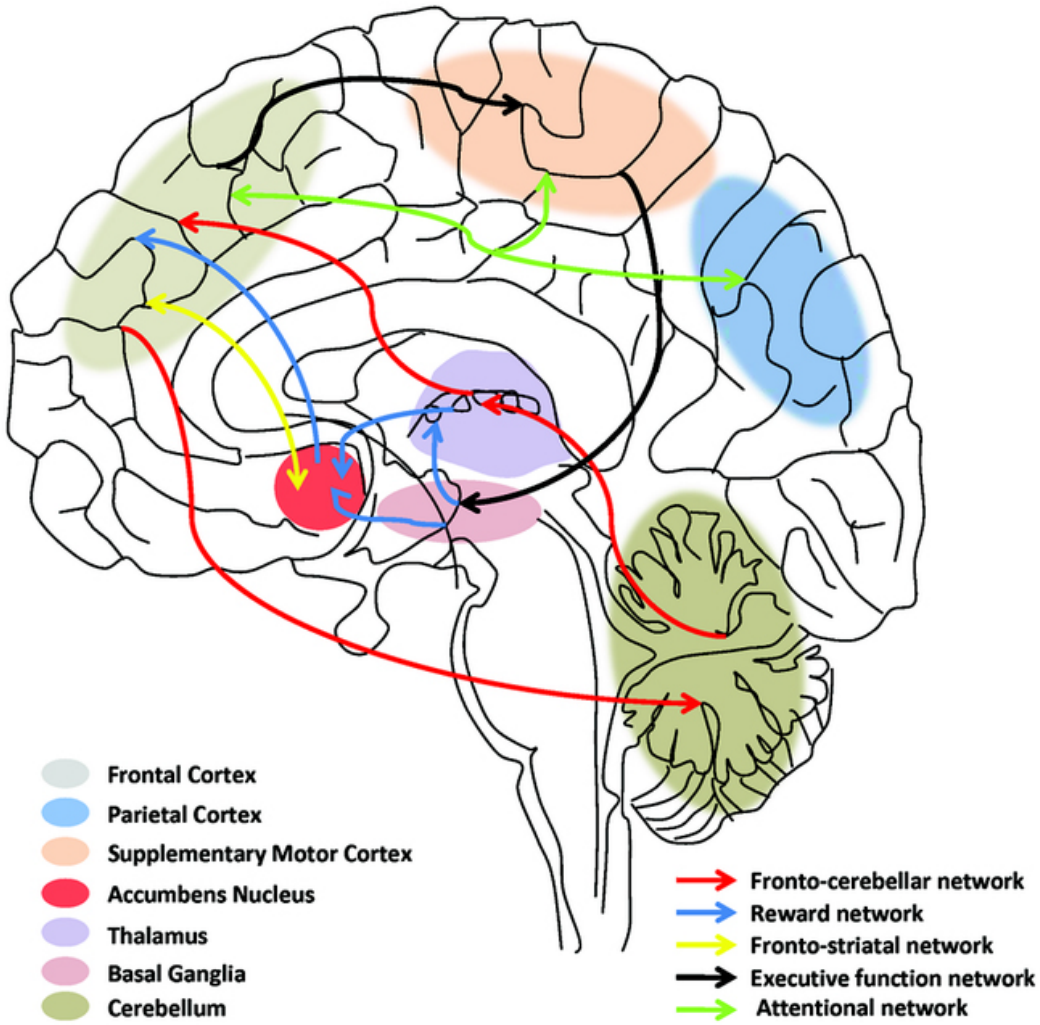
Ref: McFarland et al 2016 Armstrong et al 2013, Juten et al 2014, Wilson et al 2021

Cortical Regions



Parietal
Frontal

Cognition- subcortical and WMT



- Connectivity is important to remember
- Note how many project to frontal lobe which holds responsibility to executive functioning

Fronto-striatal
Arcuate Fasc
SLF

Cortical Basal Syndrome - language

Right and left perisylvian region

Dement Neuropsychol 2021 March;15(1):16-27

Original Article

https://doi.org/10.1590/1980-57642021dn15-010002

Language in corticobasal syndrome: a systematic review

Isabel Junqueira de Almeida[1], Marcela Lima Silagi[2], Jacy Bezerra Parmera[3], Sonia Maria Dozzi Brucki[3], Eliane Schochat[1]

ABSTRACT. Language is commonly impacted in corticobasal syndrome (CBS). However, the profile and type of language assessment in CBS are poorly studied. **Objective:** To identify language impairments in CBS. **Methods:** A search was performed in the Medline/PubMed database, according to the PRISMA criteria, using the keywords "corticobasal syndrome" OR "corticobasal degeneration" AND "language". Articles on CBS covering language assessment that were written in English were included, with no constraints on the publication date. **Results:** A total of 259 articles were found and 35 were analyzed, consisting of 531 participants. Twenty-eight studies showed heterogeneous language deficits and seven mentioned nonfluent primary progressive aphasia. The most used tests were the Western Aphasia Battery (8 studies) and the Boston Naming Test (8 studies). **Conclusion:** It was not possible to identify a unique linguistic profile in CBS.

Keywords: corticobasal syndrome, language, neurocognitive disorders, language tests.

- Central in diagnostic criteria
- Confirms "cortical" involvement
- PNFA more common
- Can be logopenic
- Perisylvian region and Arcuate Fsac
- Parietal = phonological assembly frontal = speech production
- Quite heterogeneous (disease stage, different underlying pathologies, lack of consensus of linguistic aspects to be assessed)
- Aphasic patients may progress to mutism
- Right side "emotional aspects of speech"
- PNFA may develop CBS

Cortical Basal Syndrome - Praxis

Frontal
Parietal
Basal Ganglia

- Apraxia is core feature occurring in 70-80%
- Can progress to both sides but side of onset remains worse
- Degeneration to frontal, parietal lobes and the basal ganglia result in disruption to planning, initiation and coordination of purposeful movement
- It is not a loss of desire to perform action/movement
- Ideomotor apraxia most common form (gestures impaired despite knowledge of the task e.g. comb hair). Can get eye lid and saccade apraxia
- Can be hard to assess due to bradykinesia, rigidity and limb dystonia (if present)
- It can affect dressing, preparing meals, non verbal communication, employment (if still working), risks

Wilson et al 2021

Cortical Basal Syndrome – Visuospatial

Occipital
Parietal

- Greater prevalence of visuo-spatial deficits can correlate to underlying pathology e.g. Alzheimer's pathology
- Linked to parietal lobe degeneration and involvement in “dorsal stream”
- Connectivity between subcortical and cortical area disrupts planning and integration of visual information
- Difficulty with visual processing (perceptual and constructional)
- Complicated by motor and frontal lobe involvement?

Bak et al 2006, Parmera et al 2016

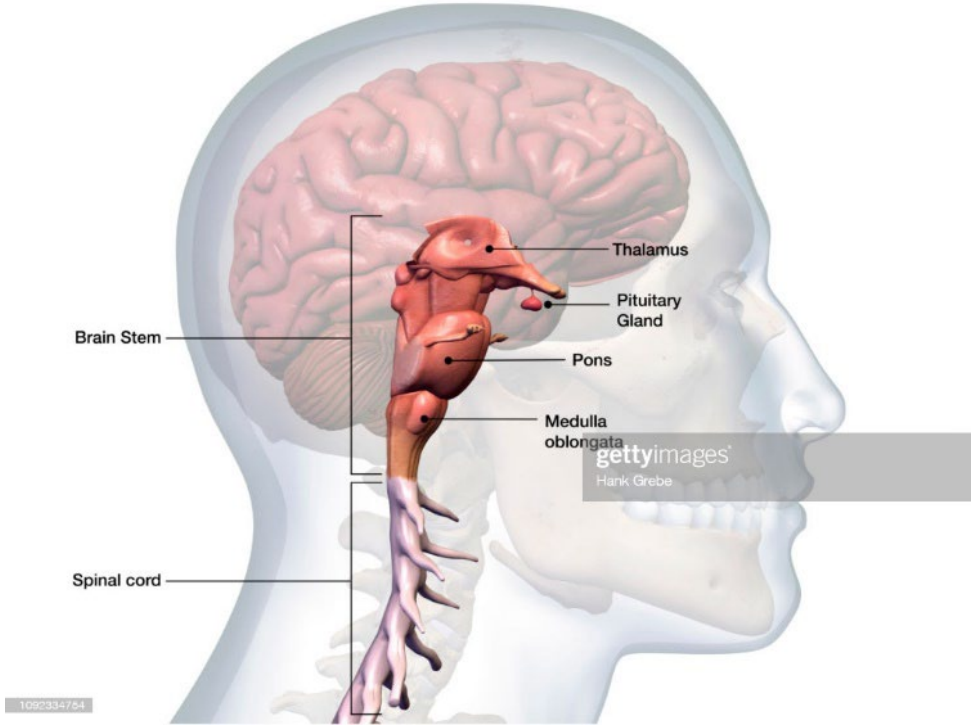
PSP

Progressive Supranuclear Palsy

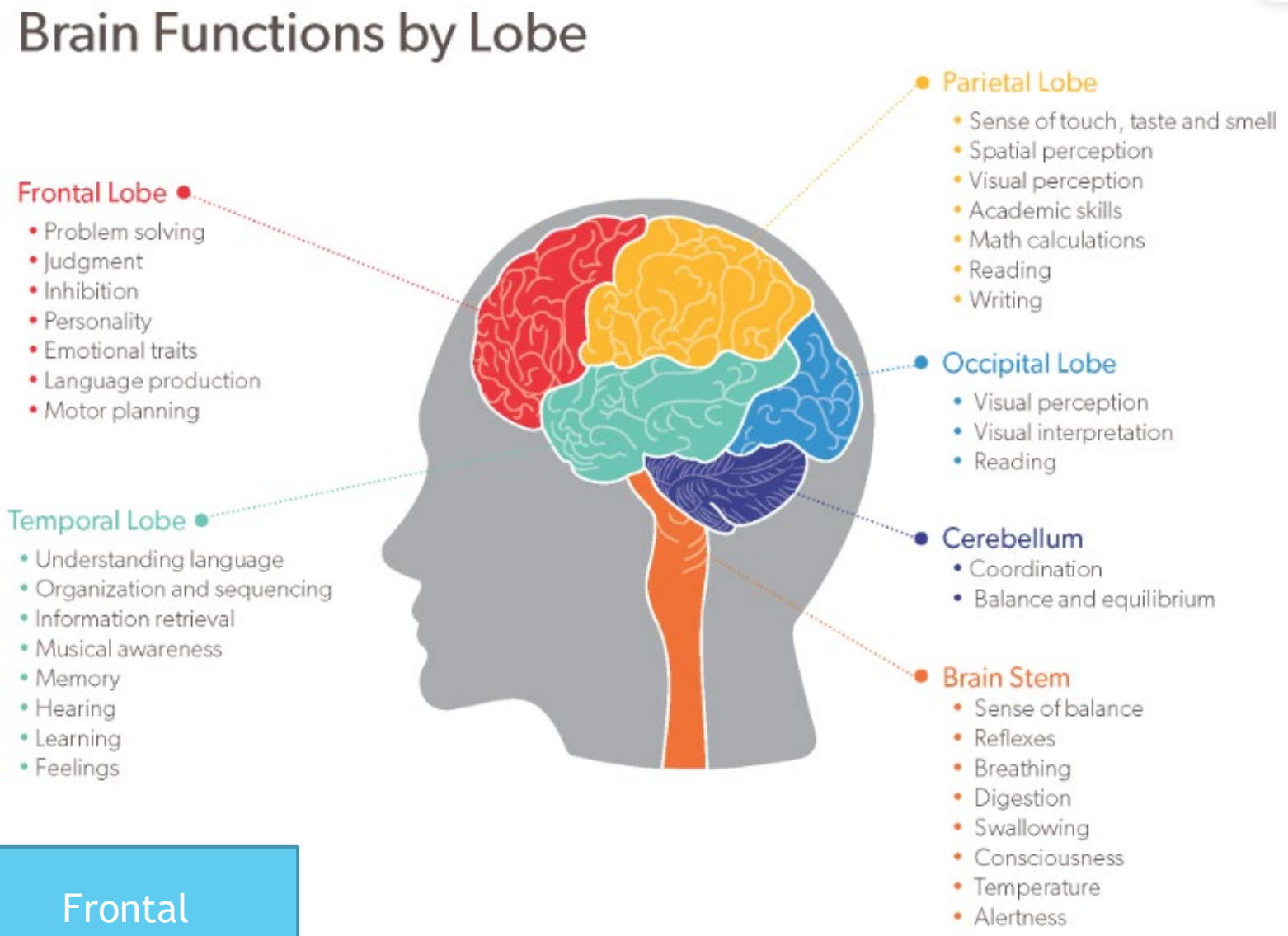
- 4R Tau deposition in nerve cells
- Midbrain, basal ganglia and frontal cortex atrophy
- Vertical gaze palsy thought to relate to atrophy of midbrain
- Postural instability and early falls (often backwards) key features
- Different phenotypes most common Richardson variant
- MRI – atrophy of midbrain tegmentum, dilation of 3rd ventricle.
- Earlier bulbar symptoms and falls in 1st year linked with poorer prognosis

McFarland et al 2016

PSP – hummingbird sign

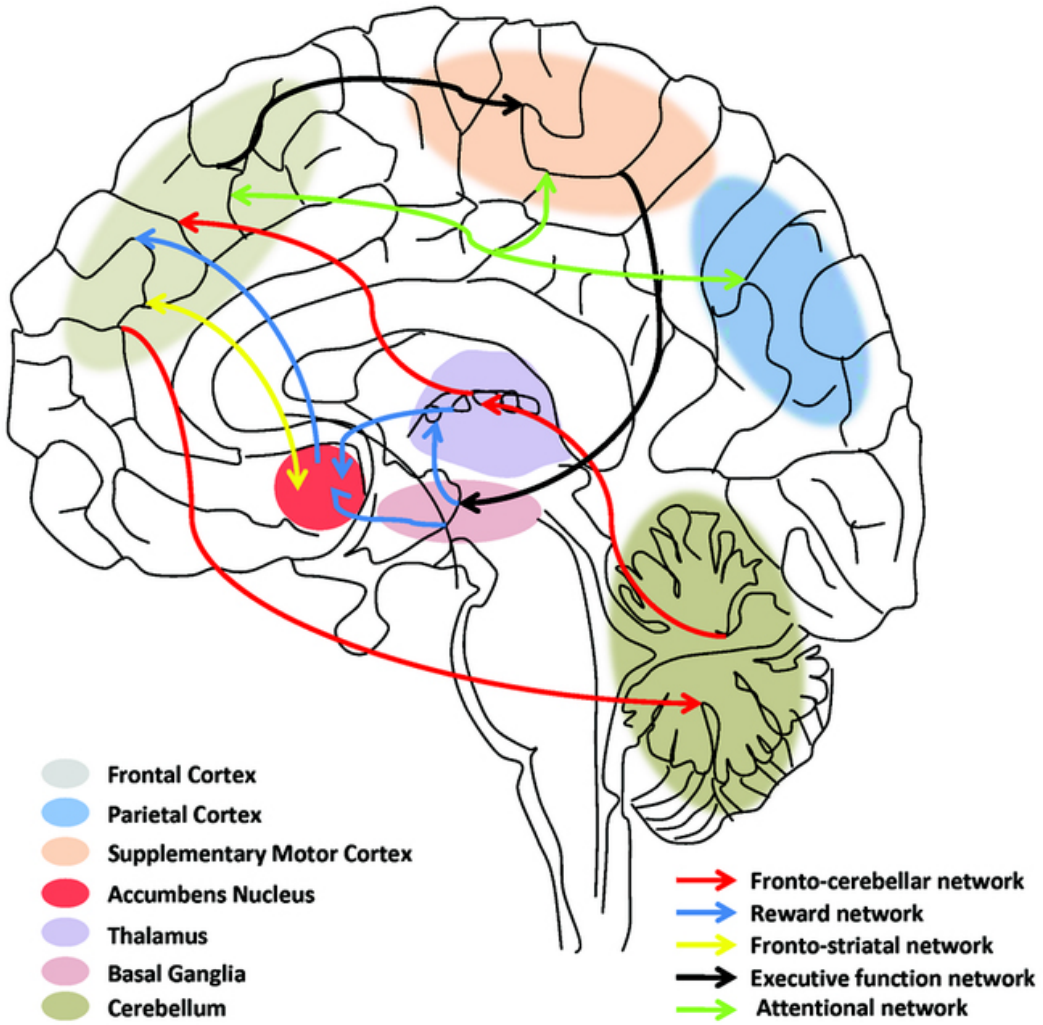


Cortical Regions



Frontal

Cognition – Subcortical and WMT



- Connectivity is important to remember
- Note how many project to frontal lobe which holds responsibility to executive functioning

Fronto-striatal (early)
SLF
Thalaomocortical

Progressive Supranuclear Palsy- Language

Frontal striatal Tegmentum of midbrain

- Different mechanisms behind speech impairment (SMC, medial frontal)
- Two main types commonly recognised in diagnostic criteria
  - 1.NonfPPA
  - 2. apraxia of speech
- Echolalic Dynamic Aphasia (not specific to PSP but can occur)
- Reduced fluency and spontaneous speech
- Dysarthria
- Pragmatics (turn taking etc)

Berthier et al 2021

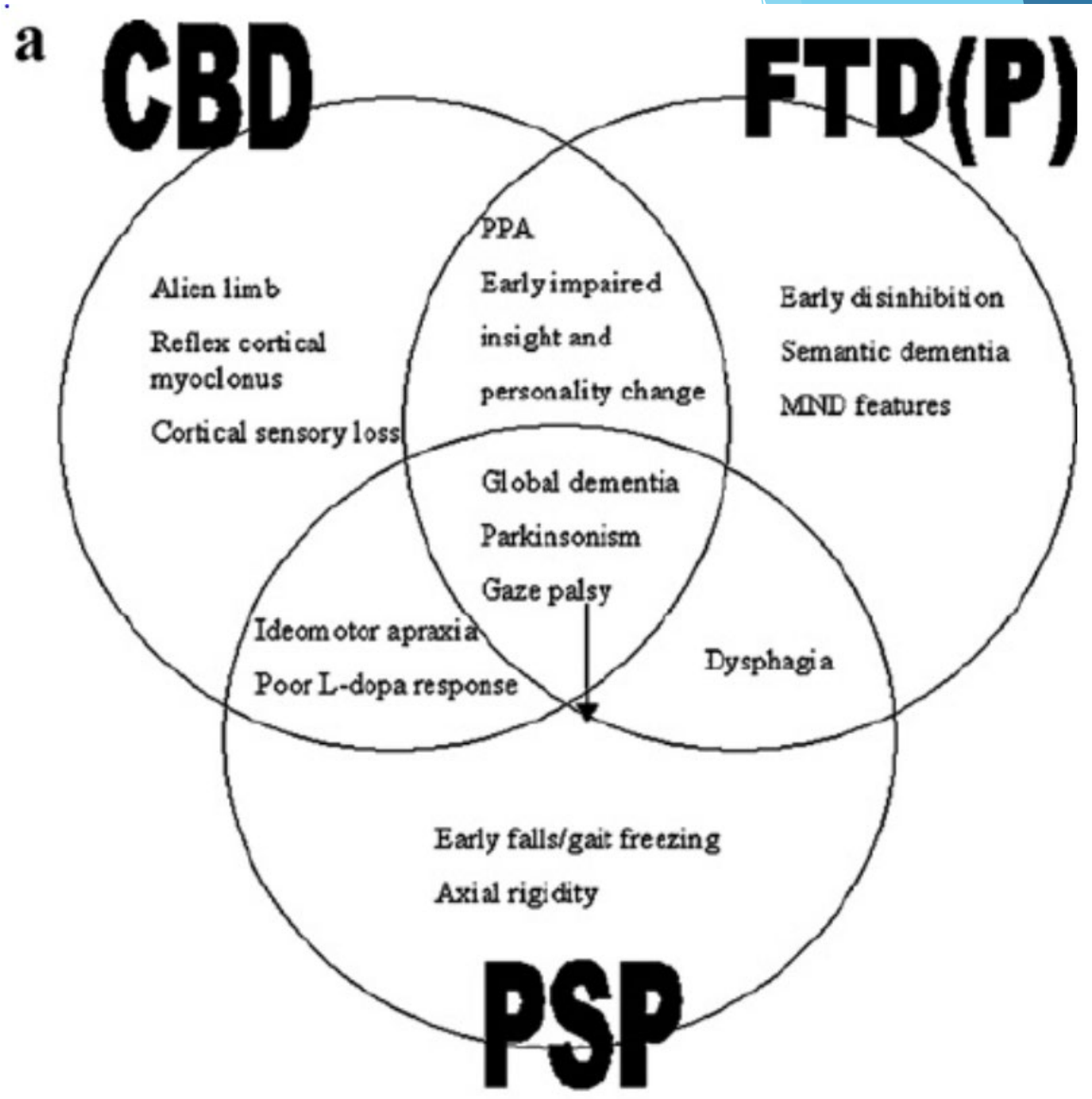
PSP –emotional expression

- Emotional lability relatively common in PSP
- PSP affects brain stem structures that hold a role in emotional regulation
- Basal ganglia significantly affected and frontal-striatal circuitry affect control of emotion
- Emotion can be disproportionate or inappropriate to the situation

Can be many phenotypes

Baseline features are often what syndromes are characterised by but this can change over time and there may be diagnostic progression in time

e.g Respondek et al 2016
76% of pathologically confirmed PSP at autopsy had a different initial clinical syndrome at presentation rather than PSP-RS



CBS & PSP – Executive Functioning

Dorso-lateral pre frontal cortex

- Very common
- Affects planning, problem solving & decision making
- Rigidity
- Initiation & inhibition
- Perseveration (seen on primitive frontal motor tasks e.g applause test)
- Significant reduced verbal fluency (worse on phonemic Vs Semantic) and worse in PSP than CBS
- Memory deficits are more a function of striatofrontal dysfunction than hippocampal atrophy e.g. better with prompts, may not be main cog issue

Ritmann et al 2016, Hu et al 2025

PSP & CBS –behaviour

Fronto striatal
Oribto- frontal
Medial frontal

- Impulsivity (PSP 54% Vs CBS 26%)
- Lowered insight (more preserved in early CBS)
- Apathy (medial frontal) – more PSP
- Reduced social cognition
- Irritability and distress (cortical-subcortical circuits affect limbic system)
- Qualitative aspects of behaviour change various
- Pattern of disruption of BG differs – PSP more caudate which inhibits movement

Ref: Hu et al 2025, Horta-Barba et a; 2021, Gerstenecker et al 2017, Ghosh et al 2012

Main differences between other neurodegeneration

- More severe and early changes than PD
- Less prominent early episodic memory impairment than Alzheimer's disease
- Less disinhibition than FTD and more apathy

Cognitive Strategies and support

Why consider this?

- Cognitive symptoms can appear early
- Drive carer giver burden
- Affects safety and independence
- Cognitive symptoms can be misunderstood or misattributed meaning the right support is not mapped on to needs
- Enhance QoL

Strategies – Executive Dysfunction

- Providing an *external frontal lobe*
- One step instruction at a time
- Errorless learning – avoids need for self correction
- Avoid open ended questions – reduces need for cognitive flexibility
- Provide choice and avoid the abstract
- E.g. X – what do you want to wear?
    - ✓ - Green or blue shirt today?
- Asking concrete questions also helps with reduced fluency which can occur due to executive retrieval deficits

Strategies – Impulsivity

- Remove trip hazards
- Use chair/bed alarms
- Position walker in front of patient
- Place visual STOP signs on walker, car door etc
- Smaller utensils, pre cut food, “one bite one swallow”
- Predictable routines – impulsivity increases when brain needs to make unstructured decisions
- Forced delay – count backwards from 3 before standing
- Environmental supports works by decreasing reliance on internal judgement
- External braking system

Strategies – Bradyphrenia and slowed processing speed

- Allow more response time (count to 10 internally)
- Do not interpret delay as refusal or non compliance
- Pause after asking a question – avoid rapid fire, this can lead to anxiety which can inadvertently increase cognitive load
- Avoid jumping in to finish sentences unless necessary
- Use visual supports to reduce cognitive load (visual reminders stay, verbal does not)

Strategies – Attention impairment

- Reduce background noise e.g. pause TV when talking, try find quiet space to talk, be aware of environment and leave important chats till later
- See this as not a courtesy but as cognitive accommodation
- Use a consistent routine where you can
- Professionally, keep sessions short
- Use written and verbal instructions (dual encoding and processing)

Strategies - Apathy and reduced initiation

- External cues as substitute for diminished internal cueing (alarms, visual prompts, Alexa reminders)
- Activity scheduling (e.g white board with a time table for the day or week)
- Routine – structured engagement in plans, hobbies etc
- Schedule rather than “wait to see if they want to”
- Don’t rely on intrinsic motivation as such – may have to take the lead
- E.g. don’t say “ do you want to go for a walk?”, say “ Its time for a walk”

Strategies – memory

- Recognition based support – cue
- Use note books, alarms, Alexa, reminders – go digital if you can!
- Be consistent with where things are placed around the house
- Label things if required so there are visual reminders and cues
- Producing cues reduces search demands on an impaired retrieval system

Strategies - Praxis

- OT colleagues helpful here
- Demonstrate – imitate – repeat
- Errorless learning – guide movement correctly
- Break steps down

Environmental changes

Environmental structure often works better than cognitive training alone. Because the disease affects internal control systems, modifying the external environment reduces the need for impaired internal regulation.

Essentially….

A helpful way to conceptualise cognitive intervention in PSP and CBS is this:

The brain's internal executive system is compromised.
Therefore, we externalize structure.

The brain's processing speed is slowed.
Therefore, we slow the environment.

The brain's initiation system is impaired.
Therefore, we initiate externally.

The brain's retrieval system is inefficient.
Therefore, we provide cues.

We are not trying to restore damaged frontal circuits. We are building scaffolding around them

Assessment

- Access may vary but Neuropsychological assessments can be helpful
  - Towards the diagnostic process
  - To help patient and family understand changes they are seeing
  - Streamline advice (mood Vs cognition or both)
  - Capacity assessments
  - Baseline
- Screening – Frontal Assessment Battery, ACE-II, MOCA (ensure training)
- More detailed assessment
  - Refer on

Mood

- Depression and anxiety can be non motor symptom
- Apathy distinct entity from Depression
- SSRI’s can be helpful but for Neurology/GP/Geriatricians to comment

Hu et al 2025
Cognitive and Neuropsychiatric profiles

- Depression and anxiety more prevalent in CBS (emotional reactivity more preserved)
- Apathy more prevalent in PSP (medial frontal lobe circuits impaired)
- Apathy correlated positively with impulsivity reflecting impairment in decisions to act or not and diffuse pathology affecting the reward control pathways

Mood

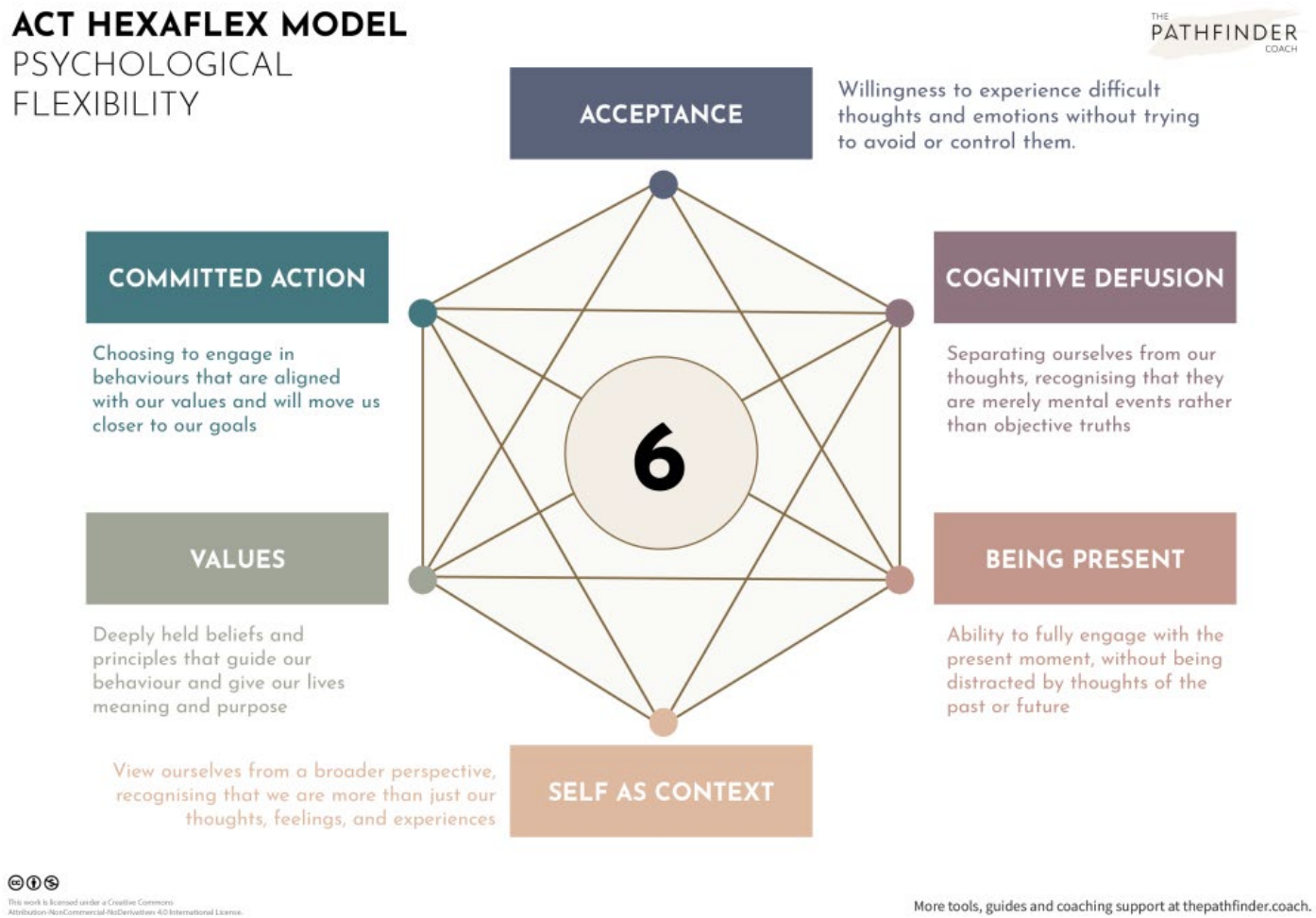
- Adjustment to living with a progressive condition
  - Grief over loss of health/relationships/roles/opportunities
  - Fear for the future
  - Fear of Burden
  - Loss of confidence
  - Systemic factors
- Mood can also affect cognition
- Is it always mood or apathy e.g. hypomimia

Treatment – limited evidence

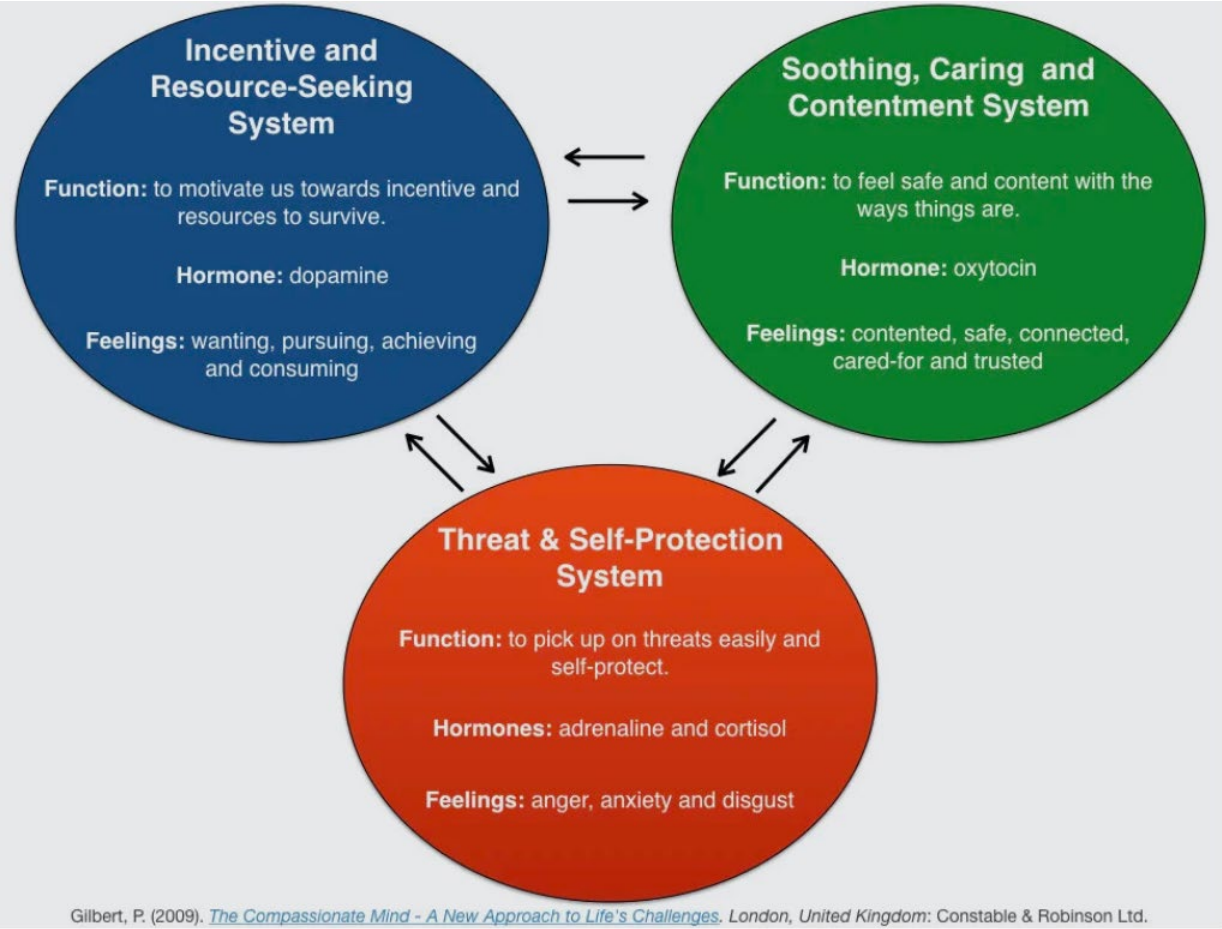
- Sparse literature on third wave (ACT, CFT) treatment
- But evidence in other physical health conditions (COMMEND trial MND)
- Clinically and theoretically, can be a better fit than CBT
- Often good approach for adjustment and values based focus
- Storey et al 2025 qualitative study of patients with neurological conditions and their experience of ACT

“stronger emotional connectedness, better knowledge of their difficulties, more effective psychological skills, and different mindsets regarding their condition”

ACT Hexaflex

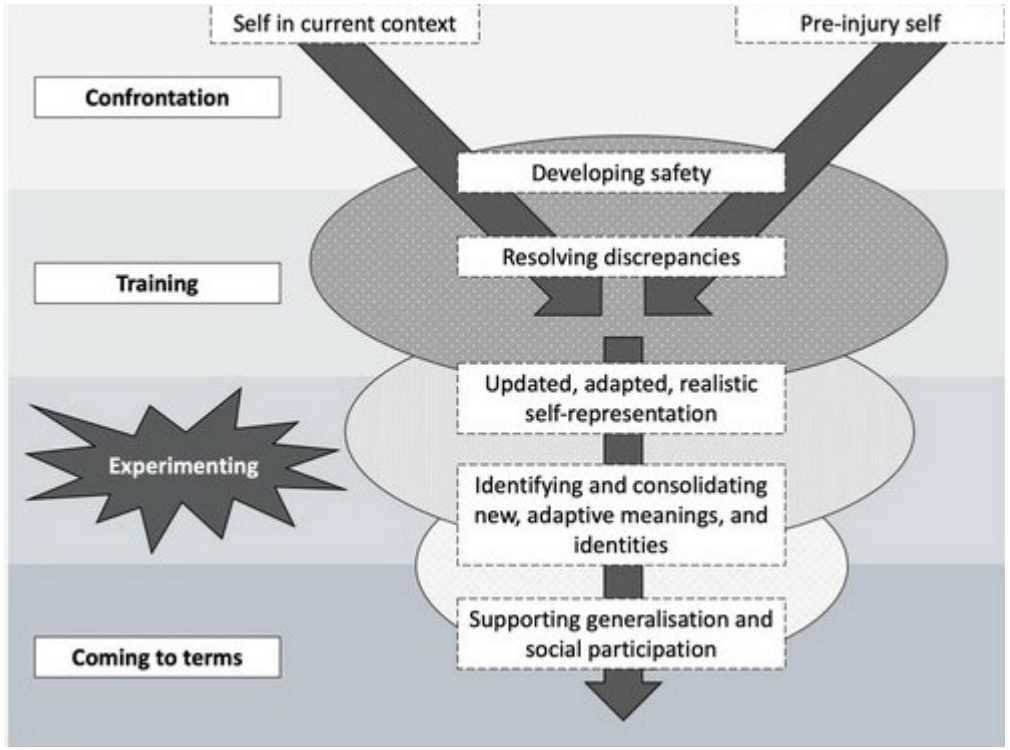


Compassion Focussed Principles



Gilbert, P. (2009). *The Compassionate Mind - A New Approach to Life's Challenges*. *London, United Kingdom*: Constable & Robinson Ltd.

Gracie Y model



Housed in CBT

But helpful model to guide conversations around sense of self and identity following diagnosis

Management & Support

- Local Services will vary
- Assessment of needs to determine which speciality can meet needs
- Distress Thermometer?
- SALT/OT/PT support
- Psychologically informed care
- Silvercloud modules? – hope for more neurological specific modules
- Mental Health Teams

It takes a team!!