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A GUIDE TO PSP & CBD
FOR PHYSIOTHERAPISTS

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This booklet is published by PSPA. It is part of a series of publications for health and social care professionals, to help them support people who are living with Progressive Supranuclear Palsy (PSP) and Corticobasal Degeneration (CBD).

This booklet aims to provide physiotherapists with information about PSP & CBD and to guide and inform their practice and intervention. Through consultation and by drawing on evidence and literature from comparable progressive neurological conditions, the guide aims to address issues at different stages of the disease progression and to provide practical suggestions to aid therapeutic intervention.

This booklet is intended as a guide only and the information is published in good faith. It is not a substitute for the advice and experience of the consultant or other medical experts involved in the provision of care.

INTRODUCTION TO PHYSIOTHERAPY IN PSP & CBD

There is limited evidence to support physiotherapy intervention in PSP & CBD, although this is changing. There is however an increasing body of evidence in Parkinson's disease (PD) and other neurodegenerative conditions and specialist palliative care services which can be transferable in many circumstances. Physiotherapy is an important part of the multidisciplinary team (MDT) who will aim to work collaboratively to help maintain independence and quality of life for those during the course of these conditions.

AS STATED IN THE CHARTERED SOCIETY OF PHYSIOTHERAPISTS (CSP) FRAMEWORK:

"Physiotherapy is a healthcare profession that works with people to identify and maximise their ability to move and function. Functional movement is a key part of what it means to be healthy. Physiotherapists recognise that physical, psychological, social and environmental factors can limit movement and function. They use their knowledge and skills to identify what is limiting an individual's movement and function, and to help individuals decide how to address their needs. The evidence-base underpinning physiotherapy is constantly evolving as practitioners develop new knowledge and understanding through critical reflection, evaluation and research. Physiotherapy works to maximise an individual's movement capability at three different levels. It can help maintain and improve the body's movement and function by offering treatment when someone is acutely ill in hospital. It can also improve someone's function and independence (at home, at work) by offering rehabilitation and advice. It can also enhance their performance and participation (in their community and wider society) by offering advice and by challenging the environmental or social barriers that limit participation."

(Physiotherapy framework 2020)

Although these conditions are rare, physiotherapists will be able to draw on their core skills and through a thorough assessment they will be able to problem solve and clinically reason to ensure that the most effective intervention is undertaken utilising the evidence available to them. The aim of this guide is to highlight the main symptoms which are specific to these conditions to help therapists guide their problem solving and intervention, ensuring a patient centred approach. Family members/significant others may also be a valuable source of information.

Figure 1: structure of physiotherapy framework showing how individual elements work together to produce physiotherapy practice CSP (2011) Physiotherapy Framework: putting physiotherapy behaviours, values, knowledge and skills into practice (updated May 2020).

MULTIDISCIPLINARY MANAGEMENT OF PSP & CBD

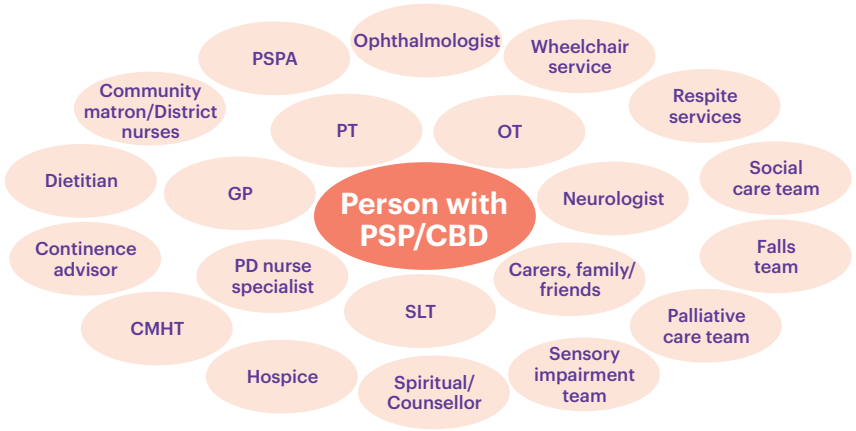
CONSIDERATIONS FOR ASSESSMENT AND INTERVENTION

PSP & CBD are rare neurological conditions with no curative treatment and an average duration from diagnosis to death of 7 years. The progression of symptoms leads to dependence on caregivers. These conditions affect speech, swallow, vision, cognition, movement, bladder and bowel function. Due to the complexity and diverse symptoms there are often many professionals involved in their care. There is often overlap in the professions but paramount is good communication between those involved to ensure that the individuals and any carers' needs are met.

We should aspire to physiotherapy provision being delivered by a therapist with specialist knowledge and skills in PSP & CBD. This may be under a neuro specific therapist or specialist community service.

Key members of the team providing support from diagnosis are shown in the diagram below.

A key worker system to trigger referrals and aid communication will help to ensure timely involvement of the appropriate professionals as the disease progresses. Even if the need for individual specialist input is negligible at the point of diagnosis, the unpredictable and possibly rapid progression of symptoms means that early contact between specialists and individuals is essential.



Key: GP General Practitioner, PT Physiotherapist, OT Occupational Therapist, SLT Speech and Language Therapist, CMHT Community Mental Health Team

Fig 2: The Multidisciplinary team for PSP & CBD (Quine, Hurford and Morton 2008)

PRESENTATION – PSP

DESCRIPTION

Progressive Supranuclear Palsy, previously known as Steele–Richardson-Olszewski Syndrome, is a neurodegenerative condition, classified as a movement disorder. It is defined by the accumulation of tau protein, which forms neurofibrillary tangles in the brain causing premature death of the neurons. The principal areas of the brain affected are:

- The basal ganglia (particularly the subthalamic nucleus, substantia nigra and globus pallidus)
- The brain stem
- The cerebral cortex
- The dentate nucleus of the cerebellum.

ONSET

Early symptoms may include:

- The inability to look up or down (known as a supranuclear vertical gaze palsy)
- Initially vertical eye movements may be slowed
- Parkinsonism ('extrapyramidal symptoms' of slowness and stiffness)
- Often symmetrical and predominant stiffness in the neck
- Falls – often backwards, but not exclusively
- Striking facial appearance often with frontalis overactivity ('startled expression') and reduced blink rate
- Cognitive dysfunction.

EPIDEMIOLOGY

- Unlikely to occur below the age of 40 – average age of onset is 62
- Median interval between onset and diagnosis is three years
- Slight male predominance in most studies.

INCIDENCE AND PREVALENCE

Prevalence: 6 per 100,000 – This constitutes around 5% of parkinsonian patients seen in the movement disorder clinic.

DIFFERENTIAL DIAGNOSIS

To date, definitive diagnosis of PSP can only be made by postmortem examination of the brain. Specialists can make the diagnosis with over 90% accuracy.

The initial symptoms can present as idiopathic Parkinson's and it may be some

time before development of the symptoms leads the neurologist or other clinician to suspect PSP. Some patients may wait two to three years before diagnosis of PSP is reached.

Cianci (2012) suggested a useful acronym to aid differentiation between Parkinson's disease and PSP – **FIGS**

F	Frequent falls, generally backwards
I	Ineffective medication, Parkinson's disease medication generally doesn't work
G	Gaze palsy
S	Speech and swallowing changes

No diagnostic tests exist but imaging techniques including SPECT, PET and MRI scans are carried out primarily as a means to exclude other neurological conditions. An MRI scan can show the presence of the 'hummingbird sign' in some people with PSP. The 'hummingbird sign' is an indication of midbrain atrophy which has been shown to help support the diagnosis of PSP.

PSP is a very individual condition, with a wide variation in symptoms and rate of progression requiring increasing care and support from the multidisciplinary team.

There are several different subtypes of PSP including, PSP-RS, PSP-PGF, PSP-P, PSP-F and PSP-CBS. Although the initial stages may present slightly differently, they will generally develop into the same phenotype of Richardson Syndrome (Hoglinger et al 2017).

RED FLAGS FOR A DIAGNOSIS OF PSP

1	**Falls**	Often backwards and without warning
2	**Postural instability**	Axial rigidity
3	**Slowness of movement**	Bradykinesia
4	**Motor recklessness**	Impulsivity
5	**Eye problems**	Restricted eye movement. May describe finding it difficult to walk downstairs due to problems with down gaze, reduced blink, double vision
6	**Speech**	Slurring of speech, soft voice
7	**Swallowing difficulties**	Liquids and solids, excessive salivia
8	**Cognitive changes**	Change in personality, irritability, apathy
9	**Emotional lability**	More easily brought to tears or laughter
10	**No presenting tremor**	

Table 1. Progressive Supranuclear Palsy diagnostic criteria

	PSP – RS	PSP – PGF	PSP – P	Others
Definite	Defined by pathology			
Probable	Vertical gaze palsy or vertical saccade slowing			
	Unprovoked falls <3 yrs or fall on pull test < 3yrs	Progressive gait freezing in 3 yrs not responsive to dopa	Akinesia and rigidity either axial or limb, dopa responsive or not	PSP – F: with frontal cognition (3 of: reduced fluency, apathy, bradyphrenia, dysexecutive, impulsivity)
Possible	Slow vertical saccades			PSP – CBS: Vertical gaze palsy or slow saccades
	> 2 steps back on pull test < 3 yrs	Progressive gait freezing < 3 yrs not dopa responsive		1 Cortical sign (sens loss, apraxia, alien limb) AND 1 MD sign (rigid, akinesia, myoclonus)
Suggestive	Macro SWJ or eyelid opening apraxia		Bradykinesia either axial or limb, dopa responsive or not	PSP – PI: Unprovoked falls <3 yrs or fall on pull test < 3yrs
	Fall on pull test or >2 steps back on pull test			PSP – CBS: 1 Cortical sign (sens loss, apraxia, alien limb) and 1 MD sign (rigid, akinesia, myoclonus)
	Fall on pull test or >2 steps back on pull test		SWJ OR EOA OR falls <3 yrs OR fall on pull < 3yrs OR Speech disorder OR frontal cognition OR Dopa resistance OR Hypokinetic dysarthria OR dysphagia OR photophobia	PSP – CBS: 1 Cortical sign (sens loss, apraxia, alien limb) and 1 MD sign (rigid, akinesia, myoclonus)

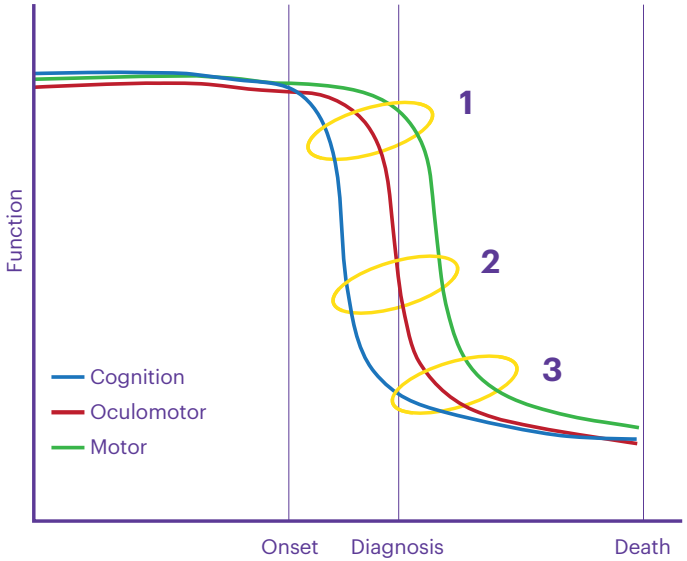
See notes overleaf

Notes

PSP-RS = PSP Richardson Syndrome
PSP-PGF = PSP progressive gait failure
PSP-P = PSP Parkinsonism
PSP-F = PSP with frontal lobe, cognitive or behavioural signs
PSP-CBS = PSP Corticobasal syndrome
PSP-PI = PSP with postural instability
SWJ = square wave jerks
EOA = eyelid opening apraxia sens loss = cortical sensory loss
MD = movement disorder
Rigid = Cogwheel rigidity
Speech disorder is non fluent aphasia or apraxia of speech.
Frontal phenotype – at least 3 of: Apathy; bradyphrenia; dysexecutive syndrome (eg. Poor Luria, stroop, reverse digit span); reduced phonemic verbal fluency; impulsivity, perseveration or disinhibition (eg. Palilalia, echolalia, applause sign, overstuffing mouth).
Mandatory inclusion: Age > 40 at first symptom, gradually progressive and sporadic.
Mandatory Exclusions: Signs suggestive of another disease e.g. MND, MSA, AD, DLB, Prion, Encephalitis; Imaging suggestive of another disease e.g. NPH; Lab findings suggestive of another disease e.g. Neurosyphilis, Wilsons, Niemann Pick disease.

PROGRESSION OF DIFFERENT ASPECTS OF PSP

Below is a diagram showing the progression of symptoms. It highlights the challenges of deterioration in eye movements, mobility and the impact of cognitive decline, particularly pertinent to this is impulsivity and the impact of motor recklessness.



An illustration of data from Ghosh et al., (2013). (Edited by Jade Donnelly)

1. Early stages of the condition, the individual may have some oculomotor changes and be experiencing instability, falls may be occurring. Therapy intervention may include, strength, core, balance and visual exercises. LSVT Big/PD warrior or a similar type intervention may be considered. Assess each individual and prescribe appropriate intervention according to their needs.

2. Mid stages of the condition, this can be a challenging time for the individual and the family. Problems with mobility can be compounded by a motor recklessness and impulsivity, which means individuals may appear to be unaware of their poor balance, and mobilise quickly and unsafely leading to multiple falls.

3. Late stages, mobility is likely to be limited and therapy may focus more on quality of life, postural management and positioning.

PRESENTATION – CBD

DESCRIPTION

Corticobasal Degeneration (CBD) is a progressive neurological disorder characterised by nerve cell loss or deterioration and atrophy of multiple areas of the brain. The progression of the disease may be slower than the progression of PSP. It is often defined by the accumulation of tau protein, which forms neurofibrillary tangles in the brain. Many of the management interventions for PSP are often helpful in CBD.

The CBD phenotype can be caused by lots of pathologies in the brain including Alzheimers, CBD or Motor Neurone Disease (MND). In this booklet we will be focusing on the CBD phenotype caused in the brain and refer to it as CBD throughout this booklet.

The principal areas of the brain affected are:

- The basal ganglia
- The cerebral cortex

People diagnosed with CBD may occasionally go on to develop features of PSP and vice versa.

ONSET

Early symptoms may include:

- The ability to use only one side of the body, for example the loss of use of one hand
- Myoclonus and apraxia (jerky, awkward movements)
- Complex unintentional movements of the limb interfering with normal tasks (alien limb)
- Increased behavioural changes
- Problems with memory
- Asymmetric – not occurring equally on both sides of the body.

EPIDEMIOLOGY

- Can affect people from 40 onwards – average age of onset 60-70
- Slightly more common in women.

INCIDENCE AND PREVALANCE

- Prevalence 5 per 100,000

Table 2. Corticobasal Degeneration diagnostic criteria

<table>
<tr><th>Probable CBD</th><th>Possible CBD</th><th>FBSS *</th><th>NAV of PPA *</th><th>PSPS *</th></tr>
<tr><td>Age ≥ 50,
<2 relatives with CBD and asymmetric presentation of 2 of:</td><td>Asymmetric presentation of 1 of:</td><td rowspan="4">2 of: Executive dysfunction
Behavioural or personality change
Visuospatial deficits</td><td rowspan="4">Effortful agrammatic speech with 1 of: Impaired sentence but preserved word comprehension
Groping distorted speech production (apraxia of speech)</td><td rowspan="4">3 of: Axial or symmetric limb rigidity or akinesia
Postural instability or falls
Urinary incontinence
Behavioural changes
Slow or palsy of vertical saccades</td></tr>
<tr><td colspan="2">a) Limb rigidity or akinesia
b) Limb dystonia
c) Limb myoclonus</td></tr>
<tr><td>2 of:</td><td>1 of:</td></tr>
<tr><td colspan="2">d) Orobuccal or limb apraxia
e) Cortical sensory deficit
f) Alien limb phenomena</td></tr>
</table>

Presentation needs to be insidious with a minimum duration of one year.

* Permitted phenotypes for Probable CBD is Probable CBD or FBSS or NAV WITH one CBD feature (a-f).

* Permitted phenotypes for Possible CBD is Possible CBD, FBSS or NAV, or PSPS WITH one CBD feature (b-f). Exclusion criteria is evidence of other diseases eg MSA, Amyotrophic Lateral Sclerosis, Lewy Body Disease, structural lesion, genetic cause or alternative primary progressive aphasia.

FBSS – frontal behavioural-spatial syndrome; NAV of PPA – Non fluent or agrammatic variant of primary progressive aphasia; PSPS – PSP syndrome.

RED FLAGS FOR A DIAGNOSIS OF CBD

1	**Highly asymmetric progressive presentation**	One side affected much earlier and worse than the other
2	**Apraxia**	Clumsy, awkward hands
3	**Dystonia**	Odd posture of hand, foot, arm or leg
4	**Myoclonus**	Quick involuntary jerks
5	**Alien limb**	Reaching or grasping automatically
6	**Speech**	Slurring or distortion of speech, halting, stuttering
7	**Cognitive and behavioural changes**	Change in personality, irritability, apathy, low mood, difficulties with organisation and planning

AETIOLOGY AND TREATMENT

DIFFERENTIAL DIAGNOSIS

Differential diagnosis is as PSP.

FAMILIAL

Less than 1% of those with PSP have a family member with the same condition. A variant in the gene for tau protein called the H1 haplotype, located on chromosome 1, has been linked to PSP, but this genetic variant is common and is not enough to cause PSP & CBD on its own.

The role of genetics in PSP & CBD is currently under investigation, but the likelihood of the condition being passed on through genetic mutations is very small.

CAUSES

Despite the recent advances in the understanding of the biology of these conditions, the cause of PSP & CBD is still unknown.

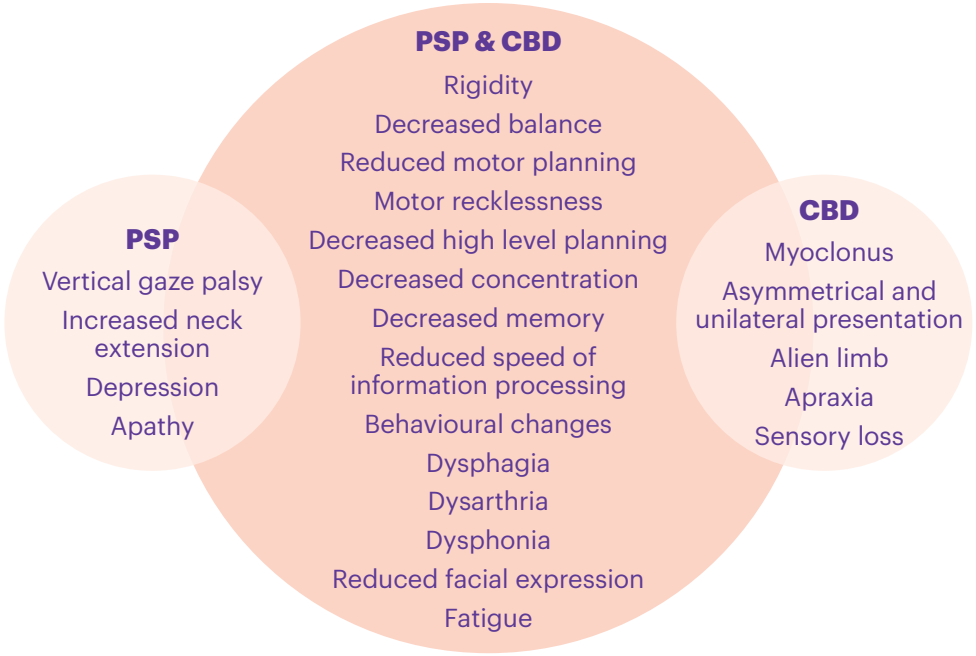
Almost all cases appear to be sporadic. It has been suggested that both environmental and genetic influences may be involved. Around 20% of the UK population carry a gene which provides a weak susceptibility to PSP (although with a very low level of risk), but the disease itself appears to be triggered environmentally or selectively.

TREATMENT

There is currently no treatment for PSP or CBD. Management is based around symptom control and quality of life hence early diagnosis and referral to specialist multidisciplinary teams is key.

IMPAIRMENTS

Impairments in PSP & CBD occur and progress at different stages and rates, with the combination and intensity varying for each individual. Some of the impairments are common in both conditions and some are specific to each one. Below are common impairments to be aware of:



Common impairments in PSP & CBD (Hurford, Morton and Quine 2008). Data from Burn and Lee, 2002 and Welling et al 1998

Symptoms become increasingly severe as the disease progresses and referral to the local specialist palliative care team should be considered early on in the disease process to facilitate more specialist support.

As with other progressive neurological conditions, people's reactions to a new diagnosis can vary significantly. The individual may experience a variety of emotions ranging from fear of the future, anger, denial and hopelessness through to acceptance. Due to the complex nature of the condition, a definitive diagnosis is often delayed. Symptoms may already be affecting the individual's performance and a degree of acceptance and adjustment may have already begun. The reactions, emotions and coping of those around them will also affect the person's response to their diagnosis.

DIFFERENCES BETWEEN DIFFERENT SYNDROMES

	IPD	PSP	CBD
Tremor	Rest	X	X
Bradykinesia	✓	✓	✓
Rigidity	✓	✓	✓
Unstable	✓	✓✓	X
Symmetrical	X	✓	X
Cognitive	X	✓	✓
Response to l-dopa	✓	X	X
Dystonia	X	X	✓
Disease duration	++	+	+
MRI findings	X	Midbrain	X

X signifies this symptom or sign is not present, increasing number of ✓s indicates increasing severity of symptoms or signs. In the MRI section midbrain changes refer to the hummingbird sign and a midbrain to pontine ratio of < 0.52 (Massey et al., 2013). MRI findings do not allow a definitive diagnosis and can be found in other conditions or not found at all. As such they provide support to the clinical impression only.

(Adapted by Jade Donnelly)

PHYSIOTHERAPY INTERVENTION

Physiotherapists working with individuals with PSP & CBD do not need to be experts in the area to provide advice on the condition and its management. It is however important the physiotherapist to have a basic understanding of the types of symptoms that are likely to be experienced through the progression of the disease and the time frames in which these may occur. In understanding this, it will enable them to judge where the individual is considered to be within this process and plan intervention accordingly. However, whilst it may be useful to plan ahead for the individual's future needs, this can often be difficult for the individual or family to accept. Be sensitive in how this is approached and also recognise that in the early stages the amount of information and the number of health and social care professionals potentially involved can be overwhelming for the individual and the family.

Physiotherapists should provide support and information to individuals living with both conditions so they can make informed decisions about their care and treatment.

The PSPA Interactive resource tool is worth looking at. The resource was developed to help drive up standards of care for those living with PSP & CBD. It provides professionals with immediate access to the very latest evidence-based information direct to their tablet or computer https://hscpguide.com/

PSPA is in contact with several professionals who have a vast body of knowledge to help support these patients and will be able to facilitate you contacting them if you feel that this would be helpful in the management of those under your care. They also provide teaching at their annual conference.

When newly diagnosed, the individual and their carers often have limited knowledge of health, social care and charitable resources available to them. At this stage, they should be directed to PSPA, which is a key resource in providing information and support.

CONSIDERATIONS FOR ASSESSMENT AND INTERVENTION

- To help strengthen your initial assessment gather as much background information as possible.
- Prior to the assessment it would be worth considering what time of the day best suits the individual in terms of their fatigue.
- Consider key impairments in relation to activity and participation in all areas of daily living.
- Acknowledge and address the carer and family's needs within the assessment process.
- Be mindful of the rate of disease progression and how this will influence current and future interventions.
- Allow sufficient time to discuss any difficulties, taking into account their exercise tolerance, communication impairments and cognitive processing skills.
- Be sensitive to their needs, returning to complete the assessment later if required.
- Consider the individual's insight into their current strengths and weaknesses and how this may affect their safety. They may have an unrealistic view of their abilities.
- Consider the individual's attitude to their diagnosis and acceptance of assistive equipment and services.
- Establish a list of main concerns and prioritise treatment goals.
- Complete appropriate referrals to colleagues from other disciplines and the voluntary sector.
- Utilise the PSPA Helpline and Information Service for further support and information for the individual and their carers.
- Arrange regular reviews of their needs and ensure that they and their carer/family know how to contact the physiotherapy service if they have any difficulties.
- Bear in mind that carers are unlikely to call for support before they have reached crisis point, which can make interventions more complex and urgent.
- Consider the individual and their carer's current knowledge of the condition and its likely course and tailor initial advice or intervention to this level.
- Patients' perceived problems.

HISTORY TAKING

- Stage of condition and current status (when were they diagnosed, severity and nature of symptoms, last neurology appointment).
- Which community services are they already known to?
- Impairments (influence of fatigue, time of day)
  - Transfers
  - Posture
  - Balance
  - Gait
- Physical activity levels.
- Falls risk.
- Comorbidities (osteoporosis, MSK disorders impacting on mobility).
- Treatment (previous physiotherapy input, type and outcome).
- Other factors:
  - Personal (insight into the condition, coping strategies, problem solving strategies, socio-cultural background)
  - Mental (cognition, mood, concentration and attention levels)
  - External factors (support networks, relationships, accommodation, work)
  - Patient's expectations (prognosis, goals and course of treatment).

SYMPTOM MANAGEMENT

VISION

Vertical gaze palsy is a diagnostic criteria for PSP. Other impairments include blurred or double vision (diplopia), reduced blink rate and tearing, photosensitivity, interrupted smooth tracking and steady gaze and reduced ability for the eyes to converge.

Gaze Palsy Patients with PSP develop a gaze palsy. This limits their ability to look down, initially movements may be slower but as the condition progresses the range will be limited and potentially paralysed. Horizontal movements may also be affected.

Assess both visual pursuits (tracking/following a target) and saccades (between 2 points) both horizontally and vertically. If there is a limitation of vertical gaze, then the vestibulo-ocular reflex (VOR) or dolls head manoeuvre can be carried out, although neck rigidity may prevent this. This would show full range of movements of the eye in the vertical direction confirming that the paresis is a supranuclear deficit, rather than a structural limitation of eye movement, which can sometimes be found in the elderly.

If patients have slowing of movements but range is preserved in the earlier and mid stages of the condition you may wish to consider incorporating scanning exercises into therapy. There is some evidence to support combining eye movements with balance exercises in the early stages (Zampieri 2008). As the condition progresses this may have limited benefit and is more likely to cause frustration.

It may be more beneficial to focus on compensatory strategies such as periscope glasses when seated to aid functional tasks such as eating/reading. These should never be worn when mobilising. Patients should be encouraged to use their neck movements, whenever possible, rather than relying on their eye movement.

Neck movements can become restricted, so it is useful to focus on cervical range of movement (ROM) early on in the condition. Patients may need prompting to scan their environment when mobilising. Remove rugs and obstacles. Steps or doorways could be marked with bright tape.

Prism/periscope glasses are available free of charge from the PSPA helpline.

Photophobia (light sensitivity causing eye closure) can have a marked impact on daily function. Drawing curtains, wearing dark wrap around glasses and a wide brimmed hat when outdoors should all be considered. Consider the environment that you are working in and the impact that this could have on the individual.

Blepharospasm (spontaneous eye closure) can also impact on function. Botulinum toxin may be beneficial. For eyelid apraxia the use of eye crutches/ ptosis crutches or Lundi Loops could also be considered. Individuals may use 'trick movements' such as tapping or applying gentle pressure to the eyebrow to aid eye opening. There is often an assumption that the patient is sleeping.

Diplopia (double vision) is also a common feature. The use of an eye patch, alternating which eye it is used on may help with this symptom. An ophthalmologist can also prescribe prisms to aid with realignment and reduce the diplopia. These prisms are different from the ones mentioned earlier which act like a periscope.

It is important to consider all of these attributes of the condition and how they might impact the individual under your care. If bifocals are used, it may also be worth advising to switch to two sets of glasses especially if they have a gaze palsy. Written material may need to be in large print and consider where you place this for the patient to best see it.

Diplopia glasses

Prism glasses

Eye crutches

MOBILITY PROBLEMS

PSP damages the regions of the brain associated with balance control, often leading to slowness in movement, motor recklessness -particularly in sitting and standing up, muscle rigidity, and deterioration in posture, gait and stamina. However, despite the rapidly progressive nature of the condition, rehabilitation and education may help to maintain balance functions and slow the decline in mobility.

Problems with mobility can be compounded by a motor recklessness and impulsivity, which means individuals may appear to be unaware of their poor balance and mobilise quickly and unsafely. This can be a very challenging symptom of the condition to manage and there isn't always an easy solution.

If the individual has problems with vertical gaze palsy, (a common problem with PSP) then they will be unable to look down and scan for hazards as they mobilise thus increasing the risk of falls further.

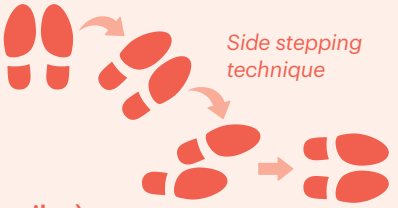
Freezing of gait (FOG)

FOG, "a brief, episodic absence or marked reduction of forward progression of the feet despite the intention to walk" (Bloem et al 2004.) PSP-PGF phenotype predominately experience FOG. (Rezvanion 2020).

This can occur when walking over a change of surface or through doorways and when they initiate gait. Optimising medication, exploring visual and auditory cueing strategies eg laser cane/laser frame with or without metronome and cognitive training could be considered.

TOP TIPS: FOG

1. Stop.
2. Stay calm, try to relax.
3. Weight transfer laterally.
4. Step back to initiate a step forwards.
5. Explore visual cues (laser beam/lines on tiles).
6. Explore tactile cues (tapping lateral thigh).
7. Explore side stepping technique if freezing when turning.



Side stepping technique

Walking aids

Physiotherapists are experts at assessing mobility and suitability of mobility aids. It is essential that an assessment includes the functional implications of mobility within the individuals different environments and tasks in order to optimise safety.

In addition, physiotherapists can provide advice on appropriate exercises to maintain and maximise function and reduce the risk of further complications. Consider a referral for rehabilitation and education if required.

- Early assessment for mobility, stairs and any appropriate aid should be undertaken and reviewed as the condition changes.
- People with PSP & CBD may benefit from a walking aid to aid balance. This ideally needs to be reviewed as the conditions change and be reassessed on an ongoing basis. When prescribing walking aids consider the stage of the condition, the environment and cognitive ability. To carry items a kitchen trolley may be considered. This may need to be adapted to include a braking system.

There is currently no primary evidence to support the effectiveness of weighted walking frames and their provision will need to be considered on an individual basis. These are not yet routinely available through statutory services and a special order or funding may need to be obtained.

- Some mobility aids have additional features such as a laser or sound cueing aid, or tension (speed) adjustment can be useful. Sticks and shoe attachments with laser cues may also be considered.
- Individuals with CBD present with unilateral and asymmetrical onset of impairments that have different functional implications for mobility compared to PSP. People with CBD may require mobility aids that only require the use of one hand and arm. Consider a stick, quad stick, or additional grab rails around the home. The 'U Step' rollator can have the additional feature of gutter handles, or a push down braking mechanism which may be helpful to individuals with asymmetrical upper limb function.
- Shoes with a wedge heel or using a wedge insert or an orthotist building up the shoe can help to transfer weight forwards so that the centre of mass is within the base of support. This may help those who have backwards disequilibrium. However, there is limited primary evidence to support the use of wedge shoes and caution should be exercised, considering the person's mobility and safety at times when they may not be wearing the shoes such as nighttime toileting or at the beginning of the day.
- In view of CBD related cognitive impairments/visuospatial deficits, wall fixed rails around the home environment may in some cases provide a more effective solution for mobility problems.
- Assessment within the home environment is key to the identification of appropriate solutions.
- Referrals for wheelchairs should be made in a timely manner, with anticipation of potential wait times which can be incurred. An explanation should be given about the sometimes lengthy wait for provision, and therefore the need to organise this sometime before it is required. Individuals may feel comfortable with this if the wheelchair is provided 'just in case.'
- Carers should be given advice and training on correct wheelchair use for care of their backs. This should include how to ascend/descend kerbs. A power pack for a wheelchair may be useful if a carer is struggling to push the chair, this is normally funded by the individual as it is not routinely funded by statutory services.
- Patients' needs should be reassessed as the type of wheelchair required may change as the condition progresses.

The combination of backward falls and difficulty with downward eye gaze in PSP makes stairs a particular hazard. Assessment of safety on stairs will need to be completed and recommendations made accordingly, for example, a second stair rail or major adaptations. Individuals who are impulsive will need appropriate risk assessment. Alternative solutions such as a bed being brought downstairs, a through floor lift (consider size to ensure it is future proof), one floor living or the support of a carer to supervise stair mobility would be useful to consider. Support with applying for rehousing may sometimes be required.

Often a stair lift may not be the most suitable long-term solution for the individual. Both conditions can often progress quickly to a stage where it is unsafe for the individual to transfer on and off at the top and the bottom of the stairs.

TOP TIP: In the early stages marking the edge of the stairs or prompting on compensation to flex at the neck to accommodate for gaze palsy may be helpful. Individuals may choose to take a greater risk to maintain independence on the stairs. Safety issue awareness should be handled sensitively.

FALLS

Falls are a major consequence of both conditions and become a constant part of the individual's and their carers lives. As such, it should be a priority during physiotherapy assessment and intervention. It is important to recognise the stress for carers related to the high risk of falls combined with impulsive behaviour and visual impairment.

Subjective history for falls

- Detailed history from the individual to determine the cause of the fall. If witnessed, obtain a collateral history.
- When did the fall occur?
  - What time of day?
  - What were they doing when they fell?
  - Where did they fall?
- What happened before and during the fall?
  - Any warning signs?
- Did they injure themselves?
- Which body part hit the floor first?
- What happened after the fall?
  - Were they able to get up?
  - How long did it take them?
  - How did they get help?
- How many falls and near misses have they had in the last 6 months?
  - The PSP Rating Scale (PSPRS) includes the frequency of falls.

- Is the individual fearful of falling?
  - Consider using the Falls Efficacy Scale-International (FES-I).
- Ask about bone health.
  - History of osteoporosis?
  - Discuss with doctors – bone health assessment (FRAX).

Guidance for falls prevention and management

- Early referral to falls management group/team. Consider referral to a NHS community therapy for a period of rehab, or advising about community based seated exercise programmes, zoom exercise programmes, or day centres who can support with exercise programmes, or private therapists/sports therapists to give 1:1 guidance.
- Referral and liaison with OT colleagues.
- Involve individual and carer in discussions about the balance between keeping physically active and falls prevention.
- Adopting a power stance (tandem stand) when opening doors and fridge for better stability.
- Avoid turning quickly, this may need to be prompted due to the impulsivity some patients have.
- Consider teaching to turn in a 'U' shape rather than a pivot.
- Education to the individual and carer to concentrate on one task at a time to reduce the risk of distraction when mobilising and transferring.
- Built up/wedge shoes that help shift the weight forwards with the aim of reducing falls backwards have been suggested by some people with PSP.
- Education on getting up from the floor for individual and carer within the home environment.
  - Backward chaining (see next page).
  - Appropriate equipment and demonstration of techniques.
- Advice on equipment available such as pendant alarms and telecare systems, including bed and chair alarms could be considered. There is also increasing use of smart phone and smart watches with GPS that can monitor falls.
- Appropriate care support to prevent falls, particularly where a vulnerable person lives alone.
- Onward referral for appropriate aids if getting up off the floor is challenging eg ELK, Mangar Camel, Raizer Lifting Chair. Consider the space for storage and physical abilities of those operating the equipment. Be mindful of the additional stress and responsibility that this might put on the carer and don't assume they are able to undertake this. Therefore, be sure to consider the pros and cons of provision of such equipment.
- Care line services with recovery team may be more appropriate if family or carers cannot physically assist.
- There is currently limited evidence to support the effectiveness of protective garments such as helmets and hip protectors, and these should be assessed on an individual basis.

Backward chaining

Falls prevention in the home

Education around the set-up of the home environment is essential for the individual and the carer to aid safe mobility. This should include:

- Adequate space for mobility aids and turning circles, and any additional equipment for transfers.
- Clear pathways and minimisation of trip hazards such as loose wires and rugs.
- The padding or removal of sharp edges and hard surfaces in likely fall areas, as it is unlikely that falls can be prevented totally.
- The use of static rails and grab handles within the home can be easier for individuals to use over mobility aids.
- Consider suitable well-fitting footwear.

SEATING/POSTURAL MANAGEMENT/SLEEP SYSTEMS

Postural changes occur and progress at different stages and rates for people with PSP & CBD. This can accelerate in the later stages and input from occupational therapy, nursing and care colleagues should also be sought. It is important to address posture early on to prevent secondary changes and support independence for as long as possible.

A physiotherapy exercise programme for posture and postural stability should include exercises to improve and maintain head and neck rotation, axial rotation, and core stability. As PSP & CBD progresses, the programme should be reviewed and appropriate assisted stretches and monitoring provided to maintain range of movement. Educate carers when to contact the appropriate services if they have concerns or see changes.

Postural changes can impact on:

- Mobility
- Transfers
- Manual handling for carers
- Bed mobility
- Pressure care/skin care
- Falls
- Contracture management
- Positioning for nutrition, enteral feeding.

Seating

Provide postural support for existing seating firstly, using cushions, pillows and rolled towels. Work with OTs to assess for optimum seating position. Consider a riser recliner chair, with suitable pressure relief. Specialist postural inserts for chairs may be able to be accessed through special funding, or charitable monies. As impairments increase it may be necessary to consider more specialist seating, and this may be needed early on. Specialist seating may be seen as a loss on independence, however it is important to focus on the positive impact this can have on their quality of life as it can allow improved positioning for eating and enjoying activities during the day, or a place to rest rather than returning to bed. It can also be a safer option if the person is at risk of falling from a regular armchair.

Consider if a specialist over-chair table or cantilever table can support an individual to continue eating independently in their chair.

Bed posture

Clinical reasoning to identify appropriate equipment and strategies to support optimum bed positioning and bed mobility is essential. Work jointly with OTs and nursing colleagues as required to balance the need for positioning, pressure care and promoting independence for toileting at night, rolling or reaching for drinks and call bells as needed.

Firstly, start by supporting with the individual's own pillows and cushions to maximise comfort and sleep, and prevent limb contractures. Focus on symmetry, alignment of posture, and increasing the base of support. A T-roll or wedge combined with pillows can be helpful. As a person's impairments progress consider specialist sleep support cushions with microbeads, or sleep systems which include trunk support brackets which can be helpful in supporting limbs, trunk and the head. Some equipment may require applications for special funding to be obtained.

A hospital profiling bed in conjunction with positioning can optimise bed posture and allow for safe manual handling and optimise a person's independence. Consider the type of bed ordered combined with pressure mattress, sliding sheets/systems, bed lever and positioning cushions to ensure best practice for pressure relief.

It is essential any postural advice and positioning is communicated also to carers via training, and pictorial charts which can be easily followed. It is important to suggest practical equipment that carers will be able to implement, and this will need to be reviewed due to the progressive nature of PSP & CBD.

Wheelchairs

As impairments increase, difficulties with mobility can become stressful for family and carers. A wheelchair may be a beneficial option but the decision can be seen as a loss of mobility and independence and an indication of deterioration. It is important to support them to see the positive impact on quality of life that using a wheelchair can have as it allows them to access activities and places further afield from the home and can increase safety.

Provision of powered wheelchairs should be considered with caution due to changes in eye gaze and cognition (impulsivity and bradyphrenia). Similar to driving, it is an all or nothing activity. However, following appropriate assessment with risk assessment and clinical reasoning it may be useful in the earlier stages of the condition. Ongoing advice and support and re-assessment of the individuals needs would be required.

SUMMARY OF MOBILITY INTERVENTIONS AT VARIOUS STAGES IN PSP

EARLY STAGE PSP

- Exercise prescription from GP/ Therapist
- Balance-specific intervention such as Tai Chi
- Falls management
- Large amplitude high intensity training ie PD warrior or LSVT BIG
- Strength – including core
- Flexibility – including axial rotation
- Functional based activities – rolling in bed
- Sit to stand (exaggerate hip hinge)
- Flexibility – Yoga/Pilates
- Boxing
- Tai Chi

MID STAGE PSP

- Modification maybe required to ensure safety maybe with therapy
- Falls management – backward chaining
- Strength
- Flexibility – axial rotation
- Functional movements – sit to stand/ rolling in bed management and equipment
- Tai Chi
- Assess/review gait and provision of walking aids
- Assess/review stairs
- Consider orthotics – wedge in shoes
- Wheelchair referral

LATER STAGE PSP

- Postural management
- Equipment needs/refer to OT
- Wheelchair/seating review

Dystonia v Spasticity

Dystonia is a symptom of a number of conditions and can be described as an unintentional, sustained muscle contraction causing repetitive unwanted movements and postures. Dystonia is differentiated from spasticity in that it is a co-contraction of both the agonist and antagonist muscle thought to be caused by a "circuit disorder involving the basal ganglia thalamocortical and cerebellothalamocortical pathways" (Albanese and Del Sorbo, 2016).

Spasticity (a symptom of damage to the upper motor neurone in a number of diseases), is due to the loss of inhibition in the spinal synaptic reflex causing overactivity in a particular muscle group (typically flexors of the upper limb and/ or extensors of the lower limb) and is velocity dependent.

Dystonia is a symptom that can present in the early, mid and later stages of PSP & CBD although often features as an early symptom in CBD.

Spasticity may also be present particularly in CBD. Dystonia in CBD and PSP is most prevalent in the arm although can affect any of the muscle groups including the eyes, neck, jaw and legs.

(Garagalvlia, et al., 2018, Bluette et al., 2021 and Stamelou et al. 2012).

MANAGEMENT OF THE DYSTONIC ARM

A typical clinical presentation of the dystonic arm is a pattern of adduction and flexion of the arm, forearm, wrist and MCPs and extension of the IP joints. Without monitoring and management this is likely to lead to secondary complications including:

- Reduced overall arm function
- Balance impairment
- Difficulty with feeding
- Difficulty with washing and dressing for the individual or carer
- Poor hand hygiene
- Infection
- Pressure damage to skin
- Pain.

Unfortunately, there is currently no evidence-based research to support the different strategies to manage the dystonic upper limb. However, best practice guidelines (Bluett et al, 2021) support the following strategies.

EARLY STAGES OF DISEASE PROGRESSION

In the earlier stages of PSP & CBD, whilst dystonia may be present, particularly in CBD, bilateral upper limb voluntary movement, although potentially impaired by apraxia, tremor and dystonia, may be preserved. At this stage beneficial interventions could include active and passive range of movement exercises and stretching programmes to maintain muscle length and avoid secondary complications such muscle contracture and joint stiffness eg adhesive capsulitis. Bilateral upper limb task specific practice has also been found in two case studies of patients with CBD to improve upper limb function and "limiting the effects of apraxia" *(Fusco et al, 2018)*.

Depending on the individual's level of cognition and walking ability it may also be helpful to provide gait re-education with a focus on contralateral arm swing.

Dynamic and static orthoses could also be considered. Whilst voluntary movement is preserved dynamic orthoses may be more appropriate to avoid restriction of intentional movement. These are discussed in more detail on page 31 and 32.

Oral muscle relaxant medications could also be considered including Baclofen and Benzodiazepines. However, these should be considered on an individual basis due to potential side effects. Anticholinergic medications should be considered carefully before prescription due to the potential to cause worsening cognitive and gait impairment. These can be discussed with the treating neurologist/physician.

Botulinum Toxin injections may also be considered with caution whilst the arm remains functional. Botulinum Toxin is discussed in more detail in the next section.

MID TO LATER STAGE OF DISEASE PROGRESSION

As voluntary activity in the affected limb becomes less available and dystonia potentially worsens other treatment modalities to be considered include passive stretching programmes, Botulinum Toxin therapy, static/dynamic orthoses and positioning.

The primary aims are to limit pain, contracture and pressure related to skin damage. Improved function is not to be expected as an intervention outcome.

Passive Stretching will need to be weighed up against any pain elicited by the movements performed and the benefit incurred by the individual as well as the ability and availability of carers and family members to provide support.

Botulinum Toxin Therapy involves injections directly into the dystonic muscles. The toxin blocks Ach receptors at the neuromuscular junction reducing nerve impulse transmission and hence, muscle contraction. For this reason, it should be approached with cautious clinical judgement when functional voluntary movement is preserved.

Injections may only be delivered by a qualified clinician, usually a neurologist or a specialist physiotherapist. The toxin normally takes around 7-10 days to take effect and its effect can be seen for up to three months. If the intervention is successful, it can be repeated on a three monthly basis for as long as it is beneficial.

Botulinum Toxin therapy is recommended by US best interest guidance (Bluett et al, 2021) in PSP & CBD with a caveat relating to cervical dystonia and the risks of aspiration particularly if dysphagia is already present.

Static Orthoses typically include a wrist-hand orthosis, a rigid splint designed to provide a prolonged stretch to wrist and finger extensors to reduce the risk of contracture. These are typically worn overnight and/or up to a four hour period during the day. As previously stated, they are not suitable for individuals with preserved voluntary movement. Precautions include pain tolerance of the individual during fitting and wearing of the splint, friction and skin damage caused by the splint and skill of carer/relative.

Another type of static orthosis is a palm protector. This is typically a sheepskin type of 'glove' that fits around the wrist and in the palm. It is mainly used where contracture is already present in the wrist and finger flexors and its primary aim is to prevent skin on skin contact/fingernails digging into the palm and so reduce risk of skin breakdown. Maintaining hygiene of the splints is also important to avoid the risk of infection.

Dynamic Orthoses for the upper limb are generally in the form of a lycra sleeve or glove which provides dynamic compression and sensory feedback. Whilst there is currently no research into the use of these in PSP & CBD there is some limited evidence that they can be effective in improving function and reducing tremor, muscle tone and associated pain in patients who have had a stroke, brain injury and patients with MS. It may therefore, be worth considering dynamic orthoses for patients with an upper limb tremor and dystonia during the early and mid stages of the disease when there is still functional volitional movement present. It is unclear whether there is any benefit when volitional movement is absent.

A major disadvantage of dynamic orthoses is the donning and doffing due to the necessary tight fitting of the garments. This could potentially inadvertently increase carer burden. Additionally, this type of orthoses may not be suitable for patients with autonomic problems in relation to temperature control or patients with dermatological complaints.

Positioning: it may be beneficial to use positioning aids to position the dystonic upper limb where voluntary movement is absent in order to optimise muscle length as far as is possible. When positioning the upper limb it is also important to consider the overall posture of the chair or bed bound individual to ensure adequate postural support is provided to avoid secondary complications associated with a rotated pelvis/side flexed trunk and cervical spine which in turn may also increase tone in the upper limb.

SUMMARY OF UPPER LIMB MANAGEMENT IN CBD

EARLY CBD	MID CBD	LATER STAGE CBD
Bilateral tasking	Compensation strategies	Contracture management
Muscle strengthening	Adaptive walking aids (U-Step)	Orthotics
Active ROM	Positioning	Botulinum Toxin
Active assisted stretching	Consider Botulinum Toxin	Positioning
	Orthotics	
	Passive ROM	

APRAXIA

Individuals with CBD often initially experience motor abnormalities in one limb that eventually spreads to affect all the arms and legs. Such motor abnormalities include muscle rigidity, limb dystonia as well as cortical features such as apraxia and alien limb.

Apraxia is a very complex symptom which can be described as a "disorder of motor cognition" (Cassidy, 2016) rendering the individual with difficulty in performing purposeful movements, resulting in a loss of ability to carry out skilled tasks with a hand/arm although strength and co-ordination is normal. This leads to clumsiness and then more profound problems with motor tasks.

Apraxia often manifests early in CBD (Cassidy, 2016) and is listed as a Red Flag when diagnosing CBD.

Apraxia should be included in the physiotherapy assessment, and link in with OT colleagues to give individual advice and support depending on the tasks affected.

SUMMARY OF DIFFERENT TYPES OF APRAXIA

- Apraxia is an inability to perform skilled motor tasks secondary to a disturbance in the processes of higher-level motor control.
- In ideational apraxia, the concepts of movement and intent are degraded and patients may not comprehend the appropriate use for the tool eg they may be able to recognise a toothbrush but unable to use it due to having lost the concept of the planned sequence of movement required to use it.
- In ideomotor apraxia, patients cannot convert the neural representation of an idea or goal into a precise pattern of motor activity and so make errors in the scaling, timing and orientation of movements eg unable to wave or brush hair. Meaningful gestures can also be assessed ie v for victory sign or thumbs up.
- In limb-kinetic apraxia, there is a breakdown in the fine individuated hand movements, often from a lesion in the contralateral prefrontal cortex or associated subcortical pathways; it is frequently very prominent in patients with Corticobasal Syndrome.
- These forms of apraxia can be difficult to disentangle in clinical practice; a practical solution is simply to interrogate the action production system by asking the patient to perform a wide variety of tasks.

INTERVENTIONS FOR APRAXIA

In a systematic review of eight studies in 2014 Lindsten McQueen et al concluded that task specific training is the most effective treatment approach for apraxia

in terms of function gain with one study (Geusgens et al., 2007) demonstrating transfer of treatment effect into the home environment and two studies (Geusgens et al., 2006, 2007) demonstrating long term effects of intervention at follow up.

However, as with much of the limited research into treatments for apraxia, both these studies were carried out on stroke patients, a nonprogressive condition, and it is not known if the results can be generalised to a CBD population.

There may be some benefit to be realised for some patients in compensatory task specific training linked to SMART goals.

TOP TIPS for assessment of apraxia

- **Try to avoid terminology around the different types of apraxia as this can become confusing due to varied terminology.**
- **Rule out other neurological features which could cause a similar presentation problem ie muscle weakness, spasticity, dystonia, bradykinesia, ataxia or sensory loss (Cassidy, 2016). Bear in mind some of these may co-exist but would not cause the problems associated with apraxia on their own:**
- **Assess ability to pantomime.**
- **Assess meaningful hand gestures.**
- **Assess complex, multi-step tasks.**
- **Assess impact of above in functional setting relating to patient goal.**

THE 'ALIEN LIMB' OR 'ALIEN HAND'

This is a strong indicator of CBD, but does not occur in everyone. With the 'alien hand syndrome' one or other hand may reach out and grasp hold of nearby objects. This might be furniture, or a utensil or even a carers arm. It is difficult to let go, even if asked and even if the person with CBD wants to. The hand may also drift about and feel strange. It is rarely distressing in its own right, but it can cause misunderstandings and be seen as 'deliberate' by other people even though the person with CBD cannot control or stop it.

Consider how this symptom may affect transfers, use of walking aid, and activities of daily living and provide education to carers so they can offer appropriate support.

RESPIRATORY CARE

Respiratory management plays a role throughout the course of the disease. Advice will be vital at every stage of a PSP & CBD patient's disease. In the early stages this may include advice to carry out breathing exercises to maintain lung function. As the condition progresses these breathing exercises will be helpful through to end stage where the risk of aspiration pneumonia is high and suction may be required.

THINGS TO CONSIDER

- Positioning – sitting as upright as possible particularly when eating or drinking, and regular positional changes.
- Swallowing ability – management of oral secretions, risk of chest infection. Patients may choose to eat according to their preferences whilst accepting there may be risks associated with this.
- Hydration.
- Nebulisers- if struggling with thick secretions (these are usually purchased independently by the patient).
- Breathing pattern (consider head and neck position in relation to this, retrocollis can be challenging when managing choking).

GENERAL BREATHING EXERCISES

- Active Cycle of Breathing Technique (ACBT) – relaxed breathing and thoracic expansion exercises combined with Forced Expiratory Technique (FET)/huffing.
- Positive Expiratory Pressure Devices (PEP) such as acapella or flutter can be used to help loosen secretions to make them less effortful to expectorate.

COUGH MANAGEMENT

- Coughing is often caused by swallowing problems resulting in aspiration, also mucous can be very thick and difficult to clear. Management should focus on keeping the swallow as safe as possible or mucous thinner and easier to expectorate.
- Slow-release opiates can be used for cough suppression for a persistent cough.

SECRETION MANAGEMENT

- Medication such as carbocisteine, a mucolytic, can be used to help to thin any phlegm to make it easier to expectorate.
- Medications such as atropine eye drops (off label) used orally may be considered initially, hyoscine is an option or glycopyrronium can be used to help dry up secretions. Caution may be required in patients with cognitive impairment. Refer to treating physician for guidance. Botulinum Toxin may also be considered into the parotid and submandibular glands.

- If secretions are distressing a suction machine can be used with a yankaeur catheter to help clear secretions from the back of the throat to help keep a patient comfortable. If a patient has used a suction machine regularly this can be continued, if secretions become a problem at end of life medication should be used to try to dry up secretions.
- Consider positioning, and liaising with SALT, palliative care – whichever is appropriate.

HOME VENTILATION TEAMS

- A specialised respiratory team is a good source of advice and input for patients who have more complex needs and may require equipment such as cough assist machines.

FATIGUE

SYMPTOMS OF FATIGUE

- Diminished energy disproportionate to any recent change in activity level.
- General weakness or limb heaviness.
- Diminished concentration.
- Decreased motivation to engage in usual activity.
- Insomnia or hypersomnia, sleep is unrefreshing.
- Perceived problems with short-term memory.
- Post-exertional malaise lasting several hours.

IDEAS FOR MANAGING FATIGUE

Prioritise activity – think about what a person wants to do and what they need to do	Use assistive equipment and aids to maximise energy use
Additional help for domestic and personal tasks	Reorganise the environment to save energy, things in frequent use should be easy to reach
Use a fatigue diary to assist with planning and prioritising	Plan frequent short rests throughout the day
Postural advice so activity is carried out in the most relaxed and efficient way	Do some exercise in line with physiotherapy advice/guidance
Anxiety management if anxiety is impacting on fatigue	Delegate tasks to others, educate family as to how this is helpful

IDEAS AROUND EXERCISE TO HELP MANAGE FATIGUE

Appropriate goal setting – keep goals very realistic	Use light exercise such as walking or simple exercise such as sit to stand
Choose an enjoyable activity	Keep a record of activity to help monitor progress, record any barriers and feelings, look back to reflect on progress or reassess goals
Plan exercise into the day	Yoga, Qigong, Pilates and Tai Chi can be helpful. They encourage breathing, stretching and improve balance
If exercise is impossible think about using a functional activity such as reaching for something in a cupboard or wiping a table with large circular movements	Drink plenty of fluids when exercising
Plan rest time after exercise	

MANAGING ANXIETY

WHAT IS ANXIETY?

Anxiety is a feeling of worry or unease about something, it is a normal response to dealing with an uncertain situation, however, it usually passes when the situation is over. When it starts to affect day to day life or causes distress more support may be needed to help manage anxiety.

When experiencing a degenerative disease coping with the loss of a person's role can lead to someone struggling to cope with day-to-day life and these increased levels of anxiety can be difficult to manage.

Symptoms of anxiety:

- Not being able to concentrate
- Being irritable
- Being easily distracted
- Feeling restless
- Having a constant feeling of dread.

If you are anxious you may also notice some physical symptoms such as:

- Having tense muscles
- Feeling short of breath
- Feeling dizzy
- Sweating
- Having a dry mouth
- Being unable to sleep
- Feeling tired
- Having digestive problems.

THINGS THAT CAN HELP...

Relaxation

There are various strategies to encourage relaxation, relaxation is a skill and therefore needs to be practiced to be effective. A person needs to try different techniques when they are not feeling particularly anxious and consciously recognise the feeling that the strategy elicits. Practicing and feeling more familiar with relaxation techniques will mean that they can be utilised confidently when a person is feeling anxious.

Examples to try with patients:

Simple breathing exercises – focus on breathing and taking controlled breaths in through the nose and out through the mouth.

Relaxation CD – Macmillan have a 'relax and breathe' CD which has relaxation exercises on it. It is a good idea to do this with your patient and note respiratory rate before and after the technique or ask the patient to rate their anxiety before and after doing the exercise. Encouraging a patient to recognise how they feel after such techniques is very important in encouraging them to continue to practice.

Visualisation – there are various visualisation scripts that can be used which encourage patients to imagine they are in their favourite place and think about the sounds/smells/sights.

Mindfulness – there are mindfulness scripts available online to talk through with patients.

Complementary therapy – massage and reflexology can help to manage anxiety.

Keep a diary – this can help identify triggers and also keep a record of strategies tried and how effective they were.

Using an online app or podcast can also help to manage anxiety.

Encourage patients to practice relaxation like any other skill, the more it is practiced the more effective it will be. A good starting point would be 5-15 minutes per day.

Acupuncture

It is thought that acupuncture can have an effect on stress and anxiety through stimulating the nervous system and activating the parasympathetic system to aid 'rest and digest' rather than 'fight or flight'. Acupuncture can also help to regulate chemicals in the brain, reducing serotonin levels and increasing the release of endorphins promoting overall wellbeing.

The limbic system is a group of brain structures, including the hypothalamus, hippocampus and amygdala, that work together to combine conscious and unconscious functions and regulate what the body does. The limbic system can help the body respond to a perceived threat, or fear or anger by activating the 'fight or flight' response, subsequently hormones are released to cause an increase in heart rate, blood pressure, respiratory rate and blood flow to organs and muscles. In the short term this response can be lifesaving, however, chronic

stress can cause the limbic system to be activated and persistent activation can cause harm to the body through a more long term release of the hormones. This can result in high blood pressure and damaged blood vessels and changes in appetite. Stimulation of acupuncture points, when associated 'deqi' sensations are achieved, has been shown to result in deactivation of the limbic system.

The parasympathetic nervous system can be stimulated via the vagus nerve using auricular acupuncture points to help to aid the 'rest and digest' side of the autonomic nervous system. Auricular acupuncture is easy for patients to tolerate as it can be done whilst sitting and the ear points can be accessed without patients having to remove clothing which can be tiring.

SLEEP

Sleep disturbances are common in PSP, the areas of the brain affected by PSP are the same areas that control the sleep/wake regulation system. Difficulty getting to sleep and staying asleep are common resulting in profound sleep deprivation without recuperation.

ADVICE TO PROMOTE BETTER SLEEP

- Good-quality sleep is very important and may help to relieve fatigue, as well as reduce the need to sleep during the day.
- Go to bed and get up at about the same time every day. Having a long lie-in after a sleepless night can lead to a disrupted sleep pattern.
- Gentle exercise like walking and keeping occupied with activities like reading, games or puzzles will help a patient feel naturally tired and ready for sleep.
- Get into a relaxing routine before bed. Try having a warm shower, reading or listening to soothing music. Listening to an audio book or a relaxation exercise on CD or an app can also be helpful.
- Make the bedroom a relaxing place to be in. Create an area that's dark, quiet and comfortable.
- Avoid large meals and stimulants like caffeine or cigarettes in the late evening. Try having a warm, milky drink before bed.
- Avoid alcohol within 2-3 hours of bedtime.
- Some medicines, for example, steroids, can cause sleeplessness. These would normally be taken before 2pm to try to avoid disturbing sleep.
- If you find it difficult to fall asleep or if you wake up during the night and can't get back to sleep again, get up and go to another room. Do something else, like read or listen to the radio, until you feel tired again. Avoid screens such as TV, computers or phones.
- Encourage patients to write down worries or concerns that are keeping them awake then they can be discussed with someone later.
- Avoid taking a nap during the day, try to do something else to distract from feeling tired, if a nap is needed then limit it to 30-40 mins and set an alarm to wake.
- Consider a warm bath, with relaxing oils or burning of essential oils such as lavender in a diffuser.

Mental exercises can also help a patient sleep. Below are a few examples which would usually take about 10 minutes to do:

- Try to remember the lines of a song or poem.
- Make alphabetical lists of girls' or boys' names, countries, trees or flowers.
- Relive in detail a favourite experience.
- Write a letter in your mind.
- Use a relaxation exercise *(See relaxation section)*.

TOP TIPS:

- **If you get cold at night, consider a heat pad or underlay.**
- **If you get cold at night use a high rated tog duvet rather than lots of heavy blankets.**
- **If you get hot at night, try using two thinner duvets so that you can throw one off.**
- **If you share a bed, consider using two single duvets.**
- **If you can't sleep, don't worry, try to rest.**

SUPPORTING FAMILIES AND CARERS

The NHS Long Term Plan (2019) states a commitment to identify and support carers, recognising that carers have a significantly increased risk of ill health "due to a lack of information and support, finance concerns, stress and social isolation."

The guidance also states that carers should not be left to manage emergency situations on their own and that contingency planning is key to ensure they are able to access out of hours and other relevant support services at the time they are required.

In 2014, NHS England published "The Commitments to Carers" which was informed by a national listening exercise with carers to identify what is important to them. Key themes included being recognised and respected in their role, recognising the carers needs for support, both in their role and maintaining their own health needs, flexibility and ensuring the overall family needs are taken into account.

Where possible, try to encourage and include the patient's family members in discussions and decision making, as a diagnosis of PSP or CBD affects a family unit rather than only an individual. Many relatives will have no or limited experience of caring for someone and some will adapt to the role more easily than others.

Some very practical support for a carer could be a list of names and numbers including the following information:

- Contact information – community MDT – named case manager.
- Out of hours services for support in a crisis.
- Adult social services contact details.
- A list of carer support resources, local and national.
- PSPA information.

The therapist will be involved in supporting both the patient and their support network as the condition progresses, advising on various aspects of symptom management at each interaction. Deterioration in physical symptoms and communication difficulties, along with behavioral changes, can be incredibly hard to manage and carer stress can be very high. Signposting to support services may be required. It is vital that the therapist takes into consideration the pressures on the carer and ensures that any care plans do not place demands on the carer that they are unwilling or unable to do. Thorough discussion will be required to ensure care plans meet both the patient and the carers needs. Encourage carers to maintain their own health and wellbeing and outline care support options to facilitate this.

- Encourage to access local hospice for patient and carer support through alternative therapies, psychological support, activities or groups.
- Consider day care services.
- Consider Continuing Health Care (CHC) funding checklist and when this might be appropriate.

Information on disease progression and end of life care will need to be when the patient and carer are ready to receive that information and both patient and carer should be involved in discussions around this information. Sometimes patients and carers will need to have differing levels of information and the therapist can support this through conversations individually, maintaining patient confidentiality and ensuring appropriate consent. Some carers may want to learn about the condition and engage with all support services whereas for others this may be detrimental. It is important to establish how the patient and carer want to manage the situation and support them in doing so in their chosen way.

TOP TIPS

- **Do not place demands on the carer that they are unwilling or unable to do, thorough discussion will be required to ensure care plans meet both the patient and the carers needs.**
- **Different family members may need different information at different times along the course of the condition. This may differ from the information that the individual with PSP or CBD requires.**

HOSPITAL ADMISSIONS

Hospital admissions may occur as a planned intervention or in an emergency situation. Due to PSP & CBD being rare conditions they are often not well known or understood. For both the individual and family such admissions can be a cause for concern. Below are a few tips which you may find helpful to share with the medical team.

TOP TIPS

- **If admission is planned provide information booklets from PSPA. Advise patients to carry medical alert cards from PSPA to share with professionals.**
- **Patients with PSP can be impulsive. Consider where they are on the ward and how you might limit their falls.**
- **Visual problems in particular photophobia (light sensitivity) may be exacerbated with the bright lights in hospital. Eye closure does not always mean that they are asleep!**
- **Patients may have bradyphrenia (slowness of thinking). Ensure that all members of the team are aware of this and allow the patient time to respond.**
- **Deconditioning is a risk for those in hospital. Consider how best to maintain functional ability.**

PLANNING FOR THE FUTURE

Patients are all different and approach conversations about the future differently. As a therapist you may feel able to engage/lead in these conversations, you may however, wish to seek guidance from other members of the MDT or advice can be sought from local specialist palliative care services.

ADVANCE CARE PLANNING

This involves thinking about and discussing how a patient would like to be cared for in the final stages of life. Thinking about this can be helpful for family members if they need to make decisions about care.

Advance care planning can be in the form of a general statement detailing wishes, a more detailed plan of anticipated situations or a legally binding document detailing treatments you do not want. It is good practice for these documents to be reviewed on a yearly basis and resigned and dated. There will be regional variations in how these discussions are documented but the principles remain the same. Below are some examples of the types of documents that patients may have in relation to their future and planning their care.

ADVANCE STATEMENT

An advance statement is a way that allows someone to explain the things that matter most to them about their care if they lose the ability to make their own decisions (this is referred to as losing 'mental capacity' in line with the Mental Capacity Act 2005). It sets down preferences, wishes, beliefs and values regarding their future care. It can include statements around where they would like to be cared for, where they would like to die, what is particularly important about their care and it can also cover practicalities such as future care for pets.

The purpose of an advance statement is to provide guidance to anyone who might have to make decisions in a patient's best interest if they have lost the ability to make or communicate decisions. Talking about future preferences is important. By writing an advance statement down, it can help to make things clear to family members, carers and anybody involved in a patient's care. Health professionals have a responsibility to take into account these wishes where possible and can support with documenting this.

EMERGENCY HEALTHCARE PLAN (EHCP) OR RECOMMENDED SUMMARY PLAN FOR EMERGENCY CARE AND TREATMENT (ReSPECT)

These are documents that cover possible anticipated situations which could occur due to disease, it details the potential situation and what the patient's thoughts and wishes are around it. These documents can be helpful to provide information on the focus of care, for example, if patients would prefer care to focus on comfort and quality of life rather than life sustaining treatments.

- What the patient would like to happen if they develop infection. Would they want to go to hospital for IV antibiotics if oral antibiotics had not helped, or stay at home knowing that without further treatment it could result in the end of their life?
- Would the patient go to hospital if they fell and had a fracture that was potentially fixable?
- Whether the patient has made a decision about accepting or declining treatments that have been offered eg PEG/RIG.

RESUSCITATION

Do Not Attempt Cardiopulmonary Resuscitation (DNACPR) - This is a document completed by a medical professional after a discussion with patients and family members weighing up risks and benefits for individual circumstances.

ADVANCE DECISION TO REFUSE TREATMENT (ADRT)

This is a legally binding document which is used to refuse specific medical interventions in the future. This includes life sustaining interventions such as being resuscitated if the heart stops, using a ventilator to assist breathing and interventions such as a feeding tube (PEG).

POWER OF ATTORNEY

Lasting Power of Attorney (LPA) is a legal document that allows another person, or people, to make decisions on your behalf when you are unable to make them yourself. LPA can be for property and financial affairs, and for health and welfare decisions. These documents can provide peace of mind for patients and families that someone they trust has permission to make decisions if they are not able to.

BRAIN BANK DONATION/DONATION TO MEDICAL SCIENCE

It is very important that if a patient has signed consent to donate to a brain bank or medical science that details of what actions to take after death are easily accessible and kept with any advance planning documents.

CASE STUDY – MIKE

Mike is a 72-year-old right-handed male who lives with his wife in a house with stairs. He has recently been diagnosed with PSP and referred for assessment. Symptoms first started 3 years ago when his speech and thought processes became slower. A year ago, his walking became increasingly wobbly, and he started falling backwards particularly on steps. At a similar time, his wife noted that he was coughing with his food a few times a week. 6 months ago, he developed difficulty with urinary leakage, finding it hard to get to the toilet in time. He has photophobia and is a little impulsive. He no longer drives.

He has echolalia (repetition or echoing of words or sounds that you hear someone else say). He had restricted voluntary upward saccades and slow downward and horizontal saccades. He could get up from a chair without the use of his hands but needed more than one attempt to achieve this. Sitting down was a little uncontrolled in terms of descent. His gait was unsteady, but he had no mobility aids. He is currently taking dopamine.

Following the initial assessment treatment included:

Strengthening work – sit to stand/stand to sit with cueing and preparatory work of forwards translation of the trunk in sitting to improve biomechanics and aid propulsion to aid sit to stand. A home exercise programme included core and global strengthening programme. Mike was considered safe to conduct a standing exercise programme holding onto his kitchen work surface. Other forms of exercise were discussed including local seated exercise groups. Mike has chosen to purchase a small set of floor pedals to use at home as he used to enjoy cycling.

Bed mobility – although this wasn't highlighted as a problem at the initial assessment, axial rigidity is a known complication of the condition. This was broken down, and rotational movements were emphasised. In conjunction with this, neck ROM exercises were also introduced with the same rationale. Maintaining ROM in the cervical spine also allows compensation for lack of eye movement.

Wrap around glasses were recommended and during therapy sessions, light was adjusted accordingly ie curtains closed to reduce the glare from outside.

A small heel wedge was introduced into his shoes, and this had a positive effect on his backwards displacement in standing. Mike was reluctant to have a walking aid at this time.

Safety discussions with regards to his impulsivity and the inherent risks were broached with both Mike and his wife.

Mike was assessed for falls prevention and taught backward chaining falls recovery. He is considering a pendant falls alarm. Stairs were assessed, and a second hand rail was fitted.

A discussion about Mikes armchair included considering chair raisers or a riser recliner chair in the future if required.

He was signposted to PSPA and also referred to OT and SLT for assessment. Also referred to GP for assessment of prostate in light of urinary symptoms, and the continence service for support.

The timed up and go improved following a 6/52 course of intervention at home. Mike was then encouraged to continue with the home exercise program which had been set up. He had an open referral back into the therapist.

CASE STUDY – SHEILA

Sheila is a 70-year-old right-handed female who lives with her husband in a bungalow. She has recently been referred for assessment following a diagnosis of CBD. In the last 18 months she has noticed that her left hand is clumsier and feels more stiff. She struggles with buttons and drops things. She was advised to stop driving as she was struggling to park the car and judge distances.

On examination she has dystonia in her left upper limb. When asked to copy meaningless gestures she struggled with her left hand. Similarly miming meaningful gestures such as brushing her teeth was more difficult on her left. She also had a cortical sensory loss in her left hand, when a number was drawn onto her palm she was unable decipher it (agraphesthesia).

Following the initial assessment treatment included:

Bilateral tasks and exercises focusing on range and strength in the upper limb.

She enjoyed yoga but was finding this more difficult, from working with the instructor, modifications were made and she was able to continue with her practice.

She was referred to the OT for ongoing advice and support. She was signposted to PSPA for support.

Following a course of assessment and treatment she was discharged and rereferred a further year later.

At this subsequent review she had developed contractures due to the dystonia in her upper limb and had limited functional use of her left hand. She had also developed dystonia in her left lower limb which was impacting on her gait and balance.

Treatment included:

Trial and provision of a U-step walker with gutter to enable the left UL to be supported and her to remain ambulant safely for short distances.

A hand and wrist splint was introduced to maintain range and limit secondary complications from the contractures. Education on hand hygiene.

Sheila had a Neurology review to consider medication management, and Botulinum Toxin injections.

A review of her upper and lower limb exercise programmes which were altered with the aim to monitor and prevent worsening contractures. Positioning was suggested for her upper limb and lower limbs when seated and lying.

As a result of her dystonia in her UL and LL bed mobility had become increasingly difficult. Working closely with the OT, a bed lever and WendyLett sheets were trialled. Sheila is aware she may require a profiling bed to support her in the future.

Seating and postural advice were also reviewed, and additional postural support was provided in sitting to support her trunk which had developed lateral trunk flexion. A postural insert for her current armchair was considered, however this was unsuccessful and she was assessed by an OT for new specialist seating to support her posture.

CONCLUSION

These conditions are demanding and resource heavy. Individuals contend with an array of rapidly changing debilitating impairments affecting their mobility and function in their home and within the community. This leads to life changing dependency on other family members. Each individual requires ongoing physiotherapy intervention to help address and manage the ongoing impairments as they change over the course of the condition. Timely intervention and ongoing reassessment, anticipating change and responding to problems is required.

Despite there being no current cure, there is an enormous amount that can be done to maintain the individual's quality of life for as long as possible if the MDT work collaboratively. Physiotherapists can draw on their clinical reasoning and previous clinical experience to help manage these incurable conditions.

We hope that this booklet provides an understanding of the impairments associated with these conditions and some practical guidance. Although there is limited research there are considerable resources supporting the role of PT in neurology and palliative care.

The evidence in this booklet has been drawn from expert opinion and experience and evidence from comparable progressive neurological conditions.

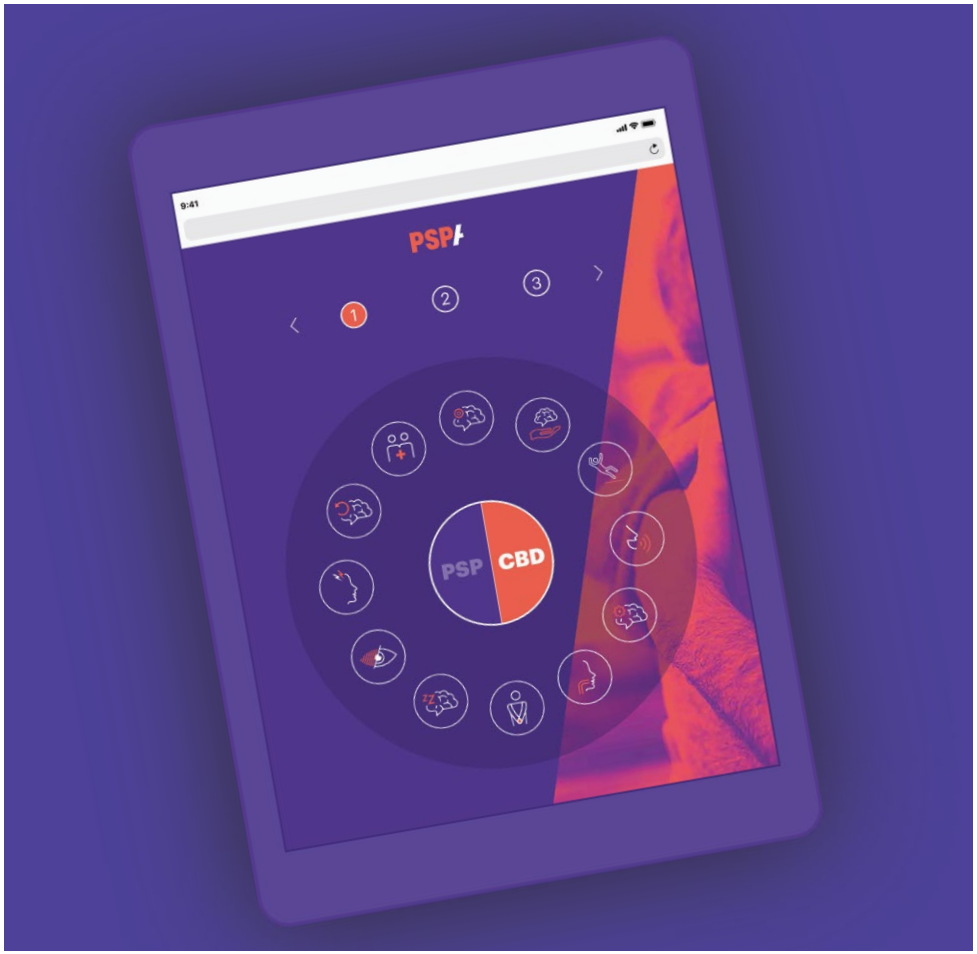
Additional information from PSPA can be found in:

A Guide to PSP & CBD for Occupational Therapists

A Professionals Guide to PSP

A Professionals Guide to CBD

INTERACTIVE RESOURCE



To provide health and social care professionals with more in depth information, an interactive resource can be found on the PSPA website for use on smartphones, tablets or desktop PC's.

www.pspassociation.org.uk

The resource gives professionals access to evidence supporting best care at different stages during the disease progression and the ability to build their own personalised guide based on the specific information they may need.

ACKNOWLEDGMENTS

A Guide to Cognition in PSP & CBD for the Primary Healthcare Team

A Guide to PSP & CBD for General Practitioners

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REFERENCES

A reference list of the evidence to support this guide is available on request. Please make your request by email to helpline@pspassociation.org.uk

Or write to us at:

Helpline and Information Services, PSPA, Margaret Powell House, 415a Midsummer Boulevard, Milton Keynes, MK9 3BN

WE WELCOME YOUR VIEWS

PSPA encourages feedback about any aspect of the information we publish. Your feedback is really important to us, as it helps us to develop new material and improve our existing information for the benefit of people living with PSP & CBD and those who care for them.

Please send your feedback to helpline@pspassociation.org.uk

PSPA

PSPA is the only national charity offering support and information to people living with PSP & CBD, while supporting research into treatments and ultimately a cure for these conditions.

PSPA offers:

- Support for people affected by PSP & CBD
- Provides information, educational resources and training opportunities for health and social care professionals
- Funds research into treatments and diagnostic tools.

PSPA Helpline and Information Service

Our Helpline and Information Service offers information and support on all aspects of PSP & CBD including symptom management, improving quality of life and signposting to other organisations.

Our telephone and email service is confidential and we aim to respond to all telephone enquires within 24 hours (during normal office hours) and email enquires within two working days. Our opening hours are Monday to Friday 9am until 9pm.

Telephones 0300 0110 122. Email helpline@pspassociation.org.uk

Information Resources

We provide high quality information resources for health and social care professionals who work with people living with PSP & CBD. We also have a wide range of resources for people affected by PSP & CBD.

Downloads of all our publications are available from our website or you can order directly from the helpline.

Support groups

We have a wide range of groups offering support to those living with PSP, CBD and their families including: Local Groups, Carers Support Groups, Newly Diagnosed Groups, CBD Group and our Youth Support Group.

If you would like more information on any of our support groups, please contact our helpline or visit our website. www.pspassociation.org.uk

Regional Support and Information

Our regional teams work to ensure that people affected by PSP & CBD have access to good local support. They are on hand to help local health and social care providers gain a greater understanding of the care needs of people and to ensure that services meet agreed standards of care. They also focus on raising the profile on both conditions by informing, influencing and educating through delivery of regional training and educational events.

Website

Our website offers accurate, timely, and relevant information on all aspects of PSP & CBD, including the latest news on research and events. www.pspassociation.org.uk

PSPA forum

Our forum HealthUnlocked is a safe place for people affected by PSP & CBD to share experiences and support each other with the challenges living with the conditions.

Our forum can be accessed via www.healthunlocked.com/psp

GLOSSARY OF TERMS

Advance care plan	Advance care planning is a process that enables individuals to make plans about their future health care. Advance care plans provide direction to healthcare professionals when a person is not able to make and/or communicate their own healthcare choices.
ADRT	An advance decision (sometimes known as an advance decision to refuse treatment, or a living will) is a decision you can make now to refuse a specific type of treatment at some time in the future.
Alien Limb	Refers to involuntary motor activity of a limb in conjunction with the feeling of estrangement from that limb.
Apraxia	Difficulty with the motor planning to perform tasks or movements when asked, provided that the request or command is understood, and the individual is willing to perform the task.
Blepharospasm	Abnormal contraction of eyelid muscles, a type of focal dystonia.
Botulinum toxin	Also known as BOTOX, is used to treat neuromuscular disorders that produce involuntary muscle contractions or spasm. The goal of the therapy is to reduce muscle spasm and pain.
Bradyphrenia	Slowed thinking and processing of information.
Brain bank	Brain banks store post-mortem brain and central nervous system tissue donated by the public for diagnosis and research into disorders. Advances in understanding genetics and many of the molecules that define brain function mean that more and more research questions can be answered from human brain tissue.
Care Plan	Defines the care and support an individual will need and who provides it. It should be documented and regularly reviewed by the care and support team in conversation with person and their family carers.

CBD	Corticobasal degeneration is a rare condition that can cause gradually worsening problems with movement, speech, memory and swallowing. CBD is caused by increasing numbers of brain cells becoming damaged or dying over time.
CBS	As there is no diagnostic test for CBD, Corticobasal syndrome may be the term used initially whilst other possible CBD mimics such as Alzheimer's and frontotemporal dementia are ruled out.
Diplopia	Double vision.
DNACPR	Do Not Attempt Cardiopulmonary Resuscitation. A DNACPR order refers only to resuscitation if the heart and breathing stops. It does not apply to or alter any other medical treatment or nursing care. A patient with a DNACPR order in their notes will still receive all of the other treatments that they might need while they are in hospital.
Dystonia	Permanent increase in muscle tone which can restrict the mobility of the affected joint and result in abnormal posture.
Legal/Lasting power of attorney	A legal document where someone (while they still have mental capacity) nominates a trusted friend or relative to look after their affairs if they lose capacity in the future.
Mental capacity	The ability to make or communicate specific decisions at the time they need to be made. To have mental capacity the person must understand the decision they need to make, why they need to make it, and the likely outcome of their decision.
Multidisciplinary care	A multidisciplinary team (MDT) is a group of health care workers who are members of different disciplines (eg doctors, nurses, therapists, social workers etc.), each providing specific services to the patient. The activities of the team are brought together using a care plan.
Photophobia	Light sensitivity causing eye closure.
Saccade	Movement of eye between two points.