



**A GUIDE TO COGNITION
IN PSP AND CBD**
FOR THE PRIMARY
HEALTHCARE TEAM



FINDING A CURE ***STARTS
HERE***

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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 you support people who are living with Progressive Supranuclear Palsy (PSP) and Corticobasal Degeneration (CBD).

PSP and CBD are sometimes presented as conditions that only affect movement and balance. However many patients and carers will have noticed changes in concentration, behaviour, or personality. Recent advances have revealed changes in the frontal and temporal areas of the brain that affect thinking and behaviour in patients with PSP and CBD.

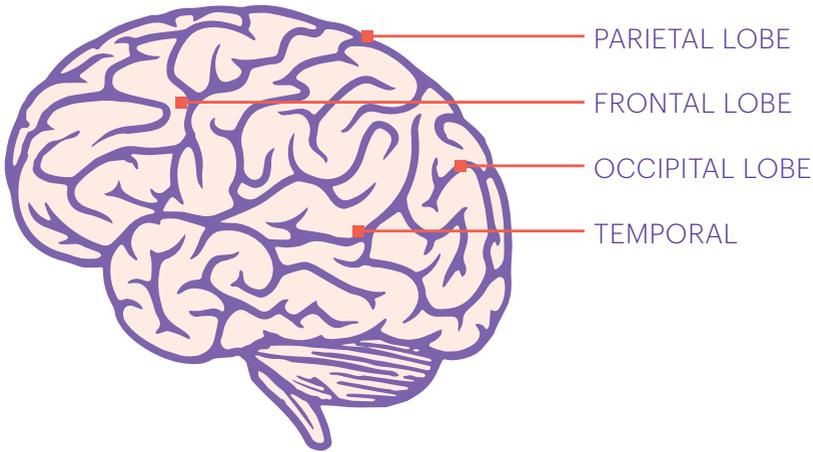
This booklet contains information on cognitive and behavioural change and dementia, and practical tips on management. It has been designed to support your work in helping people with PSP and CBD, families and carers adjust to changes in thinking and behaviour.

PSPA works in the interests of people living with PSP and CBD and often uses PSP as shorthand for both. This booklet refers to the conditions separately where there are distinct differences, as appropriate.

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

OVERVIEW OF COGNITION

'Cognition' refers to mental processes such as memory, understanding, language, vision and thinking. These use different areas of the brain, including the frontal, temporal, parietal and occipital lobes which are affected by different illnesses.



Cognition can be separated into a number of different areas:

Executive function – the skills required for problem solving, responding to new situations, shifting attention, initiating and stopping movement. Cognitive problems can cause a change in personality and behaviour. Executive functions also help us with memory.

Motivation – linked to apathy, and wanting or liking things.

Language – understanding and making written or spoken words and expressing ideas in speech or writing. Language problems are more than just difficulties with quiet or slurred speech.

Memory – learning and making new memories, storing memories and getting access to old memories when we need to. It includes memory for things that have happened and how to do things.

Vision and perception – these include information gathering by the five senses and how the brain makes sense of the information it receives to understand what one has seen, and how to look or reach for something.

Social cognition – understanding and interpreting other people’s thoughts, beliefs and feelings.

There are a wide range of cognitive changes in PSP and CBD. Some people have very mild changes, while for others they can be more pronounced, sometimes even to the point of using the term dementia.

Up to half of people with PSP have changes in cognition, behaviour or personality at the beginning of the illness, although the relevance of these changes may not be recognised until after the diagnosis of PSP is made.

When significant cognitive and behavioural changes occur early on, before problems with movement, balance, and the eyes it can look very much like a condition called frontotemporal dementia (FTD) or Pick’s disease. These are separate, although related conditions. It may be difficult to distinguish FTD from PSP. Usually people with FTD have more severe behavioural problems than those with PSP and a less severe gait or balance problem. However the two conditions can overlap or occasionally seem to change from one to the other.

Individuals with mild cognitive or behavioural symptoms will present with different kinds of problems. It is useful to discuss the presentation with the person’s care team and decide if cognitive and/or behavioural changes have been noticed and whether a formal assessment would be useful. Even mild cognitive and behavioural problems can greatly add to the stress and burden of looking after someone with PSP.

If there are significant cognitive symptoms then a formal assessment should be undertaken by a specialist with experience of PSP/CBD. A neurologist or a psychiatrist may carry out a screening assessment and may refer the person to a neuropsychologist to carry out a more detailed assessment. This helps to characterise the type and severity of different cognitive problems. Neuropsychological assessment may lead to suggestions of how to help minimise any confusion and frustration that the changes are presenting to the person living with the condition, their carers and family. Any management of changes in cognition and/or behaviour should always involve the person living with the condition, their carers and family.

For more information on how cognition is assessed in PSP and CBD (please see the *Assessing cognitive change* section in this booklet).

PRESENTATION – PSP

DESCRIPTION

Progressive Supranuclear Palsy, previously known as Steele-Richardson-Olszewski Syndrome, is a neurodegenerative condition, classified as a movement disorder. It is caused 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 or slowing of vertical eye movements)
- Parkinsonism ('extrapyramidal symptoms' of slowness and stiffness), which unlike Parkinson's disease is symmetrical and predominant in the neck rather than limbs.
- Falls – often backwards
- Striking facial appearance often with frontalis overactivity ('startled expression') and reduced blink rate
- Cognitive dysfunction including apathy, reduced speech, change in personality, mental rigidity and poor concentration.

EPIDEMIOLOGY

- Unlikely to occur below the age of 40 – average age of onset is 70
- Average time from onset to diagnosis is three years
- Average time from diagnosis to death is three to four years
- Slight male predominance in most studies.

INCIDENCE AND PREVALENCE

Prevalence of typical PSP is about 5 per 100,000. This is about 5% of parkinsonian patients seen in a movement disorder clinic. But, as many cases of PSP can present through cognitive and psychiatry services.

DIFFERENTIAL DIAGNOSIS

To date, the definitive diagnosis of PSP can only be made by post mortem examination of the brain, but specialists can make the diagnosis with over 95% accuracy.

The initial symptoms can present as idiopathic Parkinson's disease and it may be some time before development of the symptoms that lead the specialist 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

Other 'red flags' for PSP include the lack of tremor, the symmetry of parkinsonism, and early apathy.

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 may 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 be reliably predictive of PSP. But, many patients do not have the hummingbird sign when first seen and a normal scan does not rule out the diagnosis.

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.

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 also caused by the accumulation of tau protein, which forms neurofibrillary tangles in the brain but in different parts of the brain to PSP. Many of the principles of managing PSP also apply to CBD. The term Corticobasal Syndrome (CBS) is often used, but here we consider them as the same thing, to reduce confusion.

The principal areas of the brain affected by CBD are:

- The basal ganglia
- The cortex.

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

ONSET

Early symptoms may include:

- The inability to use one side of the body, for example the gradual loss of use of one hand
- Myoclonus (jerky) and apraxia (awkward or clumsy movements)
- Complex unintentional movements of a limb ('alien limb')
- Increased behavioural changes
- Problems with memory or vision
- CBD is asymmetric
- Slow, effortful speech.

EPIDEMIOLOGY

- Can affect people from 40 onwards – average age of onset 60 – 70.

INCIDENCE AND PREVALENCE

- Prevalence: about 5 per 100,000.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis is different to PSP. CBD may be mistaken for Parkinson's disease or stroke early on, and can also be mimicked by Alzheimer's disease and FTD.

AETIOLOGY AND TREATMENT

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 variation is common and is not enough to cause PSP on its own.

The role of genetics in PSP and 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 PSP and CBD the causes of these diseases are still unknown.

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

TREATMENT

There is currently no cure for PSP or CBD. But, there is a lot that can be done to reduce the severity of symptoms and seek to maintain quality of life and independence. Management is based around symptom control, quality of life and risk mitigation together with supporting autonomy and control in decision making. Hence early diagnosis and referral to specialist multidisciplinary teams is key. Later sections in this booklet discuss medication that can be offered in management of symptoms.

More information into the causes and treatment of PSP and CBD can be obtained from the PSPA website at www.pspassociation.org.uk

COMMON PROBLEMS WITH COGNITION

IN PSP AND CBD

The most common and prominent cognitive problems are apathy and poor executive function (planning and organising thoughts). Carers may find the person with PSP more irritable and more inflexible than they used to be, or they may come across as more selfish and 'unreasonable'. PSP also causes language changes, especially word finding difficulty and less spontaneous conversation. Memory is usually relatively well preserved but taking in new information can be affected by poor executive function and poor attention. Perceptual difficulties are uncommon in PSP although complaints about vision often occur, from a combination of double vision, blurred vision, sensitivity to light and glare, reduced eye movements or fatigue when reading. There can be added complications for vision in some people with prolonged eye 'blinks' lasting seconds or minutes at a time (apraxia of eyelid opening).

Sometimes a person with PSP can suddenly cry and appear to be upset for no apparent reason. This is known as emotional lability or emotional incontinence. This can be dramatic, and distressing to see, coming on and off like a light switch over a few seconds. It does not necessarily mean that the person is distressed or depressed but it can be upsetting and confusing for carers.

CBD differs from PSP in terms of which symptoms are more common, but CBD is very variable from person to person. People with CBD can also have unique cognitive problems. Visual and touch perception is often affected by CBD. It can often cause dementia with severe problems in memory and understanding language and perception, but other people with CBD have normal thinking and memory.

The most common cognitive problems in CBD are dyspraxia or apraxia. This means that there is 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 cognitive tasks. Apraxia can also affect speech, making it effortful and distorted for longer words.

A special problem in CBD is 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' or 'trouble making' by other people even though the person with CBD cannot control or stop it.

HOW DO THESE COGNITIVE PROBLEMS

AFFECT PEOPLE WITH PSP?

People with PSP may find it difficult to:

- Concentrate, eg when reading, or dealing with household bills
- Learn new activities, or use new equipment
- Hold a conversation if background distractions are present
- Do more than one thing at a time, eg multitasking or even walking and talking at the same time
- Plan ahead or manage a sequence of activities
- Make decisions with proper evaluation of choices, actions and consequences
- Start activities or tasks themselves
- Complete tasks
- Be aware of how good or bad they actually are at something and correct themselves after making an error
- Problem solve
- Find the names of items
- Know how to respond to people in social situations.

Behavioural changes that carers or family may notice are:

- Lacking interest and being apathetic
- Acting impulsively without thinking things through
- Behaving or speaking in a disinhibited manner
- Cramming food into the mouth when eating
- Preferring sweet foods
- Becoming fixated on one activity or routine
- Repetitive vocalisation, for long periods
- Indifference to the feelings of others (including carer).

If you recognise any of these symptoms perhaps in combination, discuss them with the person living with PSP, their carer and their family to see if they are aware of any changes and explore their thoughts. We recommend discussing any issues with more than one member of their healthcare team and try to identify team members who know the person living with PSP well.

IMPACT ON CARERS

Cognitive change and behavioural symptoms can cause higher levels of burden and increased stress for people with PSP, family members and carers. It is important to assess for the presence of cognitive change as early as possible after symptoms are recognised and to identify ways to help those affected.

HOW MIGHT SYMPTOMS BE EXPERIENCED AND

RECOGNISED FROM DIFFERENT PERSPECTIVES?

PEOPLE LIVING WITH THE CONDITION

Someone experiencing cognitive change may have insight in the early stages that something is wrong, but they are likely to interpret the changes as part of the physical difficulties that result from PSP or CBD. They may not recognise that their thinking or behaviour has altered.

The person living with the condition may often experience frustration or anxiety. Individuals may recognise that familiar activities they previously completed with ease are now challenging to complete. They may find it difficult to organise their activities or finances, or problem solve.

They may be aware of difficulties in finding words and understanding complex sentences, especially in crowded and busy places. They may or may not be aware that learning new tasks is difficult or that new information is more difficult to absorb or understand.

PSP can be very frustrating. But many patients, especially later in the illness, feel contented and not unwell, even though they are aware of their problems - this may be the apathy of PSP.

Depression is not common in PSP and CBD, but it needs to be considered as a possibility and treated.

FAMILY MEMBERS, FRIENDS AND CARERS

It is likely that family members and friends will see that something has changed, but there may be confusion about what the changes mean and whether they are due to the condition, or a reaction to the diagnosis or disability. They may feel concerned and protective, not wanting to broach the subject of cognitive change. They may also think that difficult or challenging behaviours are 'deliberate' and may blame or be angry with the person with PSP.

There can be relief when cognitive changes are recognised and explained, attributing the symptoms correctly to the condition. Caring for someone with poor insight into their illness can be challenging and frustrating. Despite poor mobility, balance and sight the person with PSP may not stop and think through the risks, eg getting up to walk alone. This impulsiveness and lack of awareness of danger may lead to increased risks of injury in day to day life.

They may not follow advice and this can be stressful and exhausting. It helps to understand that this is caused by changes in personality resulting from PSP, rather than stubbornness or laziness.

HEALTH PROFESSIONALS

Health professionals may notice difficulties when giving instructions or explaining procedures. They may notice apathy and lack of interest or ambivalence regarding treatments or care. Even health professionals may find it difficult to untangle the mix of symptoms and behaviour that could be interpreted at first glance as stubbornness or inflexibility.

Awareness of the type of cognitive change in PSP and lack of insight is important. It is important to rule out other causes of change in behaviour such as pain, anxiety, confusion and infections. Discuss the situation with other colleagues in the multidisciplinary team if the cause of the change in behaviour is unclear. Behavioural change like apathy or emotional incontinence may be misdiagnosed as depression, even when the person with PSP has a normal mood.

IMPACT OF COGNITIVE CHANGE CAUSED BY PSP AND CBD

Cognitive change can affect the ability to:

- Complete work, leisure and self-care activities
- Undertake new activities or learn new tasks
- Live alone without support or be left alone safely during the day
- Sustain relationships with family, friends, carers and colleagues
- Adapt to having an illness
- Make decisions about disease management including capacity to consent
- Safely operate adaptive equipment that facilitates mobility – stair lifts, electric scooters, powered wheelchairs can be hazardous with impulsive behaviour and poor reaction times
- Speech – due to vision and dexterity problems some aids may not be suitable
- Eating – people may place too much food in their mouth at one time and cram food
- Environmental controls – may be complicated and unfamiliar to learn or used unsafely

- Community alarm systems – executive function is required to realise the need to call for help and use the alarm for its intended purpose.

Symptoms may be picked up by family and professionals, but not fully understood, at various key points in the disease journey. For example, when decisions have to be made about home adaptations the person may need more time or more careful explanation to understand the need, while their reaction to the proposals may be unexpected.

For these reasons cognitive change may increase stress for people with PSP & CBD, their family and carers. It is important to assess for the presence of cognitive change as early as possible after symptoms are recognised and identify ways to help those affected.



COGNITIVE CHANGE IN PSP

MORE ABOUT FTD AND PSP

FTD and PSP are two conditions that are usually easy to distinguish. However, sometimes they appear to overlap or to slowly change from one to the other. FTD causes a wide variety of changes in behaviour and personality which are generally much more severe and occur earlier than in PSP.

The core features of the behavioural variant FTD include:

- Behavioural disinhibition, eg socially inappropriate behaviour, loss of social manners, impulsive, rash and careless actions
- Apathy and inertia, eg being withdrawn and distant, lacking interest
- Loss of sympathy and empathy
- Perseverative (repetitive), stereotyped or ritualistic behaviours and speech
- Hyperorality and dietary changes, eg sweet tooth
- Executive function and word fluency impairments, with relative sparing of memory and vision as shown in neuropsychology tests
- Lack of insight into the illness.

In addition people with typical FTD have clear changes on brain scans, such as MRI, which differ from the typical changes in PSP. The cognitive and behavioural changes in FTD can occur with limited eye movements and poor mobility or falls, causing a clinical overlap with PSP. Conversely a minority of people with PSP have severe behavioural changes that look like FTD, although these may not last. The common mild to moderate cognitive and behavioural changes in PSP do not indicate FTD. If in doubt, ask for a specialist neurological opinion.

BEHAVIOURAL CHANGES IN PSP-FTD OVERLAP CASES

This section outlines the types of behavioural changes others can expect to see in a person with overlapping PSP-FTD, which may also be called PSP-F. It represents marked behavioural change in PSP patients.

Early decline in interpersonal conduct for example:

- Apathy, a state of indifference or the reduction in emotions such as concern, excitement and motivation. An apathetic individual has an absence of interest

or concern regarding emotional, social or physical life. This needs to be distinguished from depression and fatigue in PSP

- Loss of insight
- Impaired recognition of emotions in others, or failure to understand and show concern when someone is upset or hurt
- Emotional blunting, including loss of awareness of other people's emotions and loss of insight and expression of one's own emotions.

Impaired regulation of personal conduct, for example:

- Impulsivity or acting without thinking
- Disinhibition – unable to control immediate response to a situation which may result in socially undesirable behaviour
- Continuing to engage in activity that is inappropriate to the situation
- Distractibility.

MORE ABOUT MILD COGNITIVE CHANGE IN PSP

Cognitive change in PSP commonly involves subtle deficits in executive function (problem solving, thinking through something new). Impaired word finding (verbal fluency) is very common and relates to executive function as well as language. Verbal fluency is a quick test of executive function and is an indicator of damage to frontal lobe systems.

Impairments in attention skills have also been reported in PSP. Impairments in other executive functions include:

- Distractibility
- Difficulty in sequencing and organising tasks
- Difficulty in making plans
- Difficulty in problem solving
- Difficulty with initiating ideas and thinking flexibly
- Difficulty with inhibiting and controlling thoughts.

Language dysfunction in PSP may also occur, such as:

- Reduced verbal output, eg adynamic 'aphasia'
- Perseverations, eg repetitive words or phrases
- Echolalia, eg repeating back what has been said
- Stereotypic expressions
- Difficulties understanding complicated sentences.

SOCIAL AWARENESS AND LACK OF INSIGHT

People with PSP may become less aware of what other people are thinking and feeling, and can no longer rely on facial expression or tone of voice to understand someone else's feelings and intentions. Due to loss of insight the person affected may be less aware of these changes, giving rise to misunderstandings in an already difficult situation. Carers however, may be acutely aware of the changes in behaviour and it is not uncommon to hear comments such as "they are not the same person they were". Carers may be uncertain whether the changes they are observing are caused by PSP or other causes such as depression or frustration.

For this reason it is important to rule out other causes such as fluctuations in mood, dehydration, medication or presence of infection, which can affect the persons ability to concentrate and function.

WHAT ELSE CAN CAUSE COGNITIVE CHANGE?

MOOD

The progressive nature of PSP presents a continual need for psychological adjustment. Adapting to physical problems can lead to changes in mood and many people experience frustration, anger and upset at some stage. For some the emotional change can be more profound and result in depression. Changes in concentration and memory may be related to low mood rather than PSP itself.

Most people with PSP are not depressed and many report feeling well or happy despite the many problems the illness can cause. When assessing depression in PSP it is important to focus on how the person is feeling, not how they look. Appearances can be misleading in PSP – apathy, blank facial expression and sudden crying might make the carer or professional think that the person is depressed even when they are not. However, depression is an important and treatable problem in PSP, and should be discussed. If depression occurs counselling, medication and support may help.

Psychosis is rare in PSP. Hallucinations can occur if there is an infection or in response to medication (eg Amantadine or opiate pain killers). Paranoid ideas or persistent 'odd' ideas can also occur in PSP and in CBD.

EMOTIONAL LABILITY

Some people with PSP experience 'emotional lability' which can result in sudden or uncontrollable laughter or crying. It is sometimes called 'emotional incontinence'. Sometimes this is in response to something that is moderately funny or sad (eg a television programme). There may be inappropriate responses that may be wrongly interpreted as being callous and unfeeling. This can be particularly distressing for the person and those around them but it does not mean that the person is necessarily sad and unhappy. Even when the person is not depressed certain medications (eg SSRIs) can reduce the problem and treatment should be discussed if it is causing a problem.

INFECTIONS

Chest or urine infections for example may lead to confusion or changes in behaviour, alertness, agitation and hallucinations. An indication as to whether an infection is to blame is that the change comes on suddenly, over a few days. It is sometimes accompanied by signs of infection such as fever or feeling unwell, but it is not always the case. People living with PSP may not show the common

signs of infection. For example, they may not have a raised temperature, may not cough or have a change in going to the toilet or complain of pain. If there is a rapid deterioration over the few days then infection should be considered and tested for, usually by the GP.

MEDICATION

Many medications have side effects and sometimes in PSP this can lead to cognitive change, difficult behaviours and confusion. A careful review of medication is required if cognitive change develops, asking whether each medication is required and whether it may be causing cognitive or behavioural changes as a side effect. In particular, medications used for sleep, mood, pain, bladder function and dystonia can cause confusion and even hallucinations. As the condition changes the medication list needs to be reviewed regularly.

ASSESSING COGNITIVE CHANGE

Assessment methods commonly used by clinical (neuro) psychologists, psychiatrists or neurologists include:

Interview – with the person with PSP, their family or carers to learn about symptoms, behaviour, everyday functioning, mood and experiences.

Detailed cognitive assessment – this involves a person living with PSP completing a series of pencil and paper style tests that assess their cognitive and language abilities (eg memory, planning, generating and inhibiting responses, understanding sentences and word finding). This can be a lengthy process over one or two hours, spread over appointments if necessary.

Questionnaires – people living with PSP and their carers may be asked to describe the person's behavior with questions about memory, mood, sleep, everyday activities and motivation. People living with PSP may be asked to describe their own mood in a series of questions to gain an accurate picture of psychological factors involved.

Assessment methods commonly used by occupational therapists include:

- Interview
- Functional assessment
- Standard screening of cognition and questionnaires as above.

CHALLENGES THAT MAY DELAY IDENTIFICATION OF COGNITIVE CHANGES IN PSP

Cognitive and behavioural changes are under recognised. It is easy to make the wrong assumptions about how someone is feeling or thinking. Lack of facial expression, slurred or quiet speech and physical mobility may make it hard to tell how someone is thinking or feeling. Extra problems include:

- Stigma associated with cognitive impairment and the serious implications that it has in terms of the person's ability to carry out former roles
- The subtle nature of cognitive change in people living with PSP means it can be difficult to identify within a busy clinical setting
- Lack of self-awareness and concern about cognitive and behavioural change – people living with PSP may not be motivated to report problems and may be defensive about changes reported by others

- People may attribute changes to the physical aspects of the condition, rather than difficulty with cognition eg social withdrawal may be attributed to wheelchair dependence or difficulty in speaking when it is actually caused by apathy or confusion.

THE CHALLENGES IN TERMS OF COMPLETING THE ASSESSMENT

- Motor and speech impairments often mask cognitive difficulties and make it harder and slower to carry out a proper assessment. However, it is important to give enough time to assess cognition and how the person with PSP is feeling and what they want. Often people with PSP will produce an accurate answer after a significant delay
- Time, location and resources needed to assess the aspect of functioning (particularly for those living with PSP who have subtle changes). Many teams do not have access to a clinical psychologist/neuropsychologist who is able to complete a full and detailed assessment of a person's cognitive function
- If in doubt consider referral to a specialist centre.



PROVIDING SUPPORT

Supporting carers is vital. Their needs will be individual and may be complex depending on the severity of cognitive and/or behavioural change that the person with PSP is experiencing.

This is an area that many carers feel unprepared for and understandably struggle with. Clear explanations and instruction can help. Their needs should be assessed and support strategies advised.

The need for respite is essential and may be complicated by concern for the person they are supporting and worry that other people may not understand or interpret their needs properly while the carer is absent. Communication difficulties and cognitive changes can be difficult for new staff to provide the necessary level of care, but preparation and education about PSP and the individual's needs will help.

A clear and detailed care plan is essential so that the carer feels supported and reassured that they have been listened to and that instructions are consistent and sensitively understood. Calling on family, friends and agencies that can provide support within the home may be more helpful if external respite is felt to be less appropriate.

The carer is likely to need support to explain to the person with the condition that they need a break as they may lack insight into how much the carer is doing and how exhausted they are.

Professionals must be aware of the risks to carers and families where behavioural changes include aggression. A combination of lack of empathy and self-seeking behaviour may lead to carers, family or the person themselves being in danger. Situations such as these will be challenging for the professionals, as well as carers and day-to-day management must be considered.

IS THERE A ROLE FOR MEDICATION?

- Medication can be part of the treatment strategy for cognitive problems, used together with other measures
- Medication can be used to help symptoms like anxiety, depression, poor sleep and pain that can worsen cognitive function
- Medication can help treat depression and emotional lability (SSRI's, Mirtazepine)
- Medication can help reduce agitation and restlessness

- Medication can reduce irritable, impulsive or obsessional behaviours (eg SSRI's, Trazadone - note that these are not licensed indications and should be guided by a specialist)
- In CBD, medication (cholinesterase inhibitors) can be used to help treat memory problems and hallucinations similar to their use in Alzheimers disease and Lewy Body dementias.

However, some medication can also worsen the cognition, attentional difficulties (especially anti-cholinergic drugs used for bladder symptoms or sialorrhoea) or hallucinations (amantadine, opiates). Avoid traditional neuroleptics like haloperidol as these can cause severe and sometimes irreversible extrapyramidal side effects (EPS) in people with PSP or CBD. Some antipsychotics (eg quetiapine or clozapine) are less likely to do so, but are rarely needed.

RECOMMENDATIONS FOR MANAGEMENT AND SUPPORT

The following guidelines have been added to help identify and manage what can be disturbing changes for people living with PSP, CBD and their carers.

Ways you may be able to help people with cognitive or behavioural problems:

- Remember that difficulty processing information and/or behavioural problems may be a result of cognitive change. If cognitive change is evident this may interfere with informed decision-making and use of new equipment and new routines
- Where cognitive change is present in order to communicate clearly and directly use simple straightforward language, with closed questions (that can be answered with a short simple answer), rather than open questions
- Consider whether cognitive change is causing a problem. Look at particular areas such as work, home and relationships
- People living with PSP or CBD can have varying degrees of cognitive impairment
- Consider that cognitive change may not impact on the person living with the condition as a result of changes in lifestyle. Changes at home, support from carers and occupational advice can greatly reduce the negative impact of cognitive problems on everyday life and the stress for carers.

Problems with decision making and processing complex information

- Support with the decision making process
- Break down complex information into smaller chunks

- Use clear written material rather than verbal instruction
- Take time to ensure understanding at each step
- Take time and check that there is an understanding of consequences of each action and decision
- Be prepared to wait a long time for an answer – sometimes ten or twenty seconds.

Simple decision making

- Give choices with one or two alternatives
- Do not use open ended questions. Ask questions that require yes or no answers or a simple factual response. These also work best if the person is using gestures to communicate (thumbs up/down).

Difficulty learning a new task

- Encourage the person to stop and think
- Reduce the cognitive load by breaking down the task into small steps
- If the person living with the condition fails to recognise verbal/non-verbal prompts then try to refocus their attention or show them what to do
- Suggest organisational aids, eg calenders, diaries, reminders.

Help problem solve by

- Refocusing attention on relevant issues
- Helping to monitor their own performance
- Helping to provide feedback
- Lots of practice to reinforce the steps required.

Impulsivity

- Supervise activities. People may make decisions too quickly without remembering to be careful or to use safety equipment. Encourage them to stop and think
- People may say yes when meaning no. If important, check the answer
- Supervise eating, and use smaller utensils to reduce cramming food
- Consider chair supports, height and belt if motor recklessness to 'stand-and-fall'.

Word finding difficulties (language impoverishment)

- Allow extra time for the person to find the right word
- Encourage non-verbal responding, even gestures (although this can be more difficult later in the illness)
- Try modelling the behaviour you want to encourage, eg demonstrate the task.

Passive and withdrawn

- Understand that they are likely to have difficulty initiating activities
- Use visual and verbal cues to prompt activity
- Aim for a structured routine
- Be patient and avoid irritation.

Perseveration

- Explain the problem to family and carers in terms such as ‘Mrs X has difficulty shifting her attention away from an activity once she has started. She will continue to do the same activity even though it is no longer appropriate to the situation. She may appear to be stubborn or not listening properly, but this is due to a problem in her thinking’
- Help to refocus on a new task
- Encourage a calm, structured and orderly environment.

Difficulty getting ready or organised for the day

- Focus on one activity at a time
- Engage interest and remove distractions
- Breakdown tasks into discrete steps
- Use verbal and non-verbal prompts to refocus attention or show what to do
- Minimise interference
- Plan extra time.

Change in eating habits

- Supervise the person eating more closely
- People living with the condition with more severe changes may place too much food in their mouth at one time and cram in additional food, while others may eat more than they need. Smaller spoons and forks may be useful as well as verbal prompts and gestures to slow down

- If food cravings are noticeable question how much of a problem the behaviour is causing. It may be helpful to discuss this with the dietitian.

If eating habits change or are compounded by swallowing difficulties

- Those with poor swallowing may have trouble following medical advice to modify consistency or thicken drinks
- Refer to speech and language therapist for assessment and advice on how to encourage the person to eat safely
- Raise the plate so that the person can see better. Consider using prism glasses at mealtimes
- Ensure that mealtimes are protected from any distractions.

Egocentricity (eg loss of concern for partner/carer)

- Support partner and family in understanding the reasons behind apparent selfishness by explaining that this is the person's thinking
- Explain it is part of the condition and not personal
- Encourage extra support, care or sitting service and regular respite.

Less able to respond appropriately to family or carer

- Those affected may have trouble distinguishing facial expressions
- Support partner and family in understanding the reasons behind what appears to be an inappropriate caring response
- Advise family, carers and friends they should express their feelings verbally and as simply as possible
- Recommend that they check understanding and repeat as necessary.

The person living with the condition may be less facially expressive and if behaviour is apathetic this may increase problems in recognising what the person is feeling or requires. In this situation it is important to take note of more intuitive signs of distress eg positioning, unusual/new behaviour, movement, non-verbal sounds.

Socially inappropriate behaviour (eg emotional lability, loss of control, sexually disinhibited comments)

- Explain to the carer and family that this is part of the condition, to foster understanding and support
- Explain that studies have shown that people living with PSP may have reduced understanding of emotion and social situations

- There may be misinterpretation of other people's expression eg difficulty recognising the difference between happy and surprised facial expressions
- There may be difficulty in understanding the emotions and thoughts of others
- Support partner/family with strategies to refocus/redirect attention
- Ask for specialist advice on whether medication may have a role, but avoiding difficult situations and responding with calm, consistent understanding, is the key
- Explain that the lack of concern is not deliberate or selfish but comes from a lack of understanding as a result of the illness
- Confrontation, argument and 'reasoning' is unlikely to work.

Finally, the needs of children should be kept in mind as they may need behaviour explained in age-appropriate terms they understand.

ADVANCE CARE PLANNING (ACP) AND ADVANCE DECISION TO REFUSE TREATMENT (ADRT)

Discussion of advance care planning is recommended if cognitive change is identified, however the discussion will need enough time. It is for the person with PSP or CBD to decide whether they wish to complete an ADRT. It is important to document any discussion so that their wishes are respected, particularly in the absence of any formal statements such as ADRT.

An ADRT is a decision someone can make in advance to refuse specific treatments in certain circumstances in the future. This can include the right to refuse life-sustaining treatment.

Many people living with progressive conditions such as PSP or CBD fear losing control and not being able to tell health professionals their decisions.

An ADRT tells people about those decisions and becomes active when the person loses the ability to make decisions. For many people, an ADRT can give peace of mind, enabling them to die with dignity on their own terms.

You may wish to refer people to '*Advance Decision guidance notes for people living with PSP and CBD*'.

SUMMARY

Awareness of cognitive change in PSP and CBD is improving, but for many years it has not been properly recognised or adequately treated. Professionals have too often informed people living with the conditions and their families that although PSP and CBD pose serious physical challenges, their cognition would remain intact. Research now suggests that this is not true and cognitive change is common. Dementia is unusual in typical PSP although common in CBD. Recognising cognitive change brings the opportunity to empower and educate people living with PSP and CBD.

Knowing that cognitive change is part of the condition and the disease process may come as a great relief to people living with PSP or CBD and their families who have noticed changes in thinking and behaviour since diagnosis.

If cognitive change is expected it is important that we know exactly how it effects the individual so that strategies can be put in place for appropriate care. This can help the person feel more in control and support family members and carers to understand what is happening and why. Empowering them to help the person they care for. Being alert for cognitive/behavioural change early on in the diagnosis may be valuable as this may have an impact on service use/decision making.

The management of people living with PSP and CBD who present with cognitive impairment should include selective medication, forward planning and organising appropriate support strategies for them, their families and carers.

Finally, it is important when applying any of this information to the care of a particular individual that you discuss it as a team. This allows for broad discussion and awareness from different professionals perspectives. Discussion should include the person living with the condition, and the family so that distinction can be made between normal responses in the face of changes caused by the condition and subtle yet distinct change attributable to underlying cognitive change. It also ensures that strategies suggested are applied consistently and in keeping with the individual and family routines and lifestyle.

FURTHER RESEARCH IS NEEDED IN THE FOLLOWING AREAS

New studies are required to determine how common cognitive change is, the risk factors for the individual with the condition and the rate of progression of cognitive change.

Screening tools are being developed to assess cognitive change and identify PSP-FTD or PSP-F in a clinical setting. New tools will aim to separate the difficulty in cognitive tests due to cognitive problems as opposed to physical and communication disabilities.

The root causes of PSP and CBD remain unknown. Neuropathological investigations are necessary to determine how underlying cell and protein abnormalities in the brain lead to PSP and CBD and cognitive change, as well as genetic studies to determine whether phenotypic variations in cognitive functions reflect underlying genetic differences.

Treatments for symptoms of both conditions are required including cognitive and behavioural change, these are needed in addition to treatments to slow down/ reverse the conditions.

WHO CAN HELP?

If you recognise the changes that have been described in this guide discuss the issues as a team. It is recommended that the GP, members of the hospital (neurologist and specialist nurses) and the multidisciplinary team are included. The number of people invited to team discussion will depend on the degree of cognitive change and extent of problems felt to be arising from these changes.

MILD CHANGES

People who are mildly affected it may be useful to refer to OT (for strategies and equipment to manage activities of daily living), SLT (for strategies and communication aids) or clinical (neuro) psychologist where available. Their role involves assessing understanding and ability to follow instructions. The neurologist or psychiatrist will also be able to advise on treatment of cognitive problems and how to avoid making them worse.

MODERATE TO SEVERE CHANGES

People with more severe and noticeable changes will need specialist support and advice. It may be helpful to see their OT, SLT or clinical neuropsychologist who can help identify changes which can inform the following services who may be referred to for home support:

- GP
- Palliative care team
- Community mental health team
- Cognitive neurology services/memory clinic
- Old age psychiatry services
- Local multidisciplinary team.



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

PSPA offers:

- Support for people affected by PSP and 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 and 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 5pm and then 7pm until 9pm.

Telephone: 0300 0110 122.

Email: helpline@pspassocation.org.uk

INFORMATION RESOURCES

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

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

LOCAL GROUPS

Many of our volunteer-led groups invite visiting speakers such as benefits advisers and health and social care professionals, to share expertise on managing PSP and CBD on a daily basis.

If you would like more information on our local 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 and 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 of both conditions by informing, influencing and educating through delivery of regional training and educational events.

OUR WEBSITE

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

www.pspassociation.org.uk

PSP FORUM

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

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

USEFUL RESOURCES

Rare Dementia Support

Rare Dementia Support runs specialist support services for individuals living with, or affected by, FTD and other rare dementias.

Email: contact@raredementiasupport.org

Website: www.raredementiasupport.org

Admiral Nurses

Admiral Nurses are specialist dementia nurses, working in the community with families and carers of people with dementia.

Telephone: 0800 888 6678

Email: info@dementiauk.org

Website: www.dementiauk.org

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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

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WE WELCOME YOUR VIEWS

PSPA encourages feedback about any aspect of the information we produce. Your feedback is really important to us, as it helps 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

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