

COGNITION AND BEHAVIOUR

Progressive Supranuclear Palsy (PSP) and Corticobasal Degeneration (CBD) are complex conditions. Those living with these conditions may experience a wide variety of symptoms. How these symptoms affect people differs but many people with PSP or CBD will face challenges with cognition and behaviour.

For the families of people living with a diagnosis of PSP or CBD, changes in their loved ones' thought processes, behaviour or personality can be puzzling and frustrating. This information sheet explains the cognitive changes sometimes seen in PSP and CBD and looks at strategies for coping - and how to access specialist support.

**INFORMATION STARTS
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What is cognition?

Cognition refers to mental processes such as memory, understanding and thinking. Cognitive function has several aspects including:

Executive function – the skills required for problem solving, decision making, responding to new situations and moving attention from one task to another. Executive functions also help us with memory.

Motivation – the desire to do things. It is the crucial element in setting and attaining goals.

Language – understanding and making written words or speech, expressing ideas.

Memory – learning and making new memories, storing memories, recalling memories. It includes memory for things that have happened and memory of how to do things.

Perception – the process where we take in sensory information from our environment using all our five senses and use that information in order to understand and interact with our environment.

Visual and spatial processing – the ability to understand where we are or where objects are in our environment. Also helps us recognise and understand letters and numbers, read maps and navigate.

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

How is cognition affected in PSP/CBD?

A significant number of people with PSP and CBD have changes in cognition, behaviour or personality. These changes may be evident at diagnosis or may be noticed as the condition progresses. For some people these changes may be mild whilst for others they may be more severe.

The most common cognitive and behavioral issues in PSP are:

- Apathy (lack of motivation)
- Poor ability to plan and organise thoughts
- Inflexible thinking
- Lack of empathy
- Difficulty finding words
- Emotional lability (inappropriate responses such as crying or laughing).

PSP is very variable from person to person.

The most common cognitive and behavioral issues in CBD are:

- Apraxia (difficulties in performing movements – eg difficulties in hand function – often the hand will appear clumsy)
- Perception (the way in which you think about and understand the world around you)
- Memory – CBD can cause severe difficulties with memory and understanding but many people with CBD have normal thinking and memory.

CBD is very variable from person to person.

Occasionally extremely challenging behavioral changes may occur in people with PSP or CBD. The individual may display, for example, aggression or their behavior may be socially disinhibited eg behaviour becomes inappropriate or out of character.

What else can cause cognitive change?

There can be other reasons for cognitive change in people with PSP or CBD including:

Mood – the person with PSP/CBD is constantly trying to adjust to their progressive physical symptoms and may experience frustration, anger and upset. Changes in concentration and memory can be related to low mood rather than cognitive change. For some, the emotional change can be more profound and may result in depression. If the person is struggling with low mood then it is important that the issue of depression is considered in a timely manner and, if appropriate, counselling, medication and support offered.

Emotional Lability – (sometimes referred to as ‘emotional incontinence’) can affect people with PSP/CBD and cause them to laugh or cry in an uncontrollable way. Sometimes this may seem an inappropriate response to the situation. It does not necessarily mean the person is unhappy or that they are being callous or unfeeling. Certain medications and therapeutic support may help so seek professional advice.

Infections – if there is a sudden change (often over a few days) in an individual’s cognitive ability and they become confused or agitated, or experience hallucinations, then infection should be considered as a possible cause. People living with PSP or CBD do not always experience fever or other signs of infection and they may not have a raised temperature. Chest and urine infections can be tested for by the GP and, if appropriate, treatment commenced.

Medication – many medications have side effects which can lead to cognitive change, difficult behaviours and confusion. A detailed medication review from a medical professional can help identify this.

Dehydration – people whose fluid intake is low run the risk of becoming dehydrated. This can lead to the person being confused. Ensuring the person with PSP or CBD has regular fluid intake is important.

Progression of cognitive issues

Cognitive difficulties in PSP and CBD are usually progressive. However the progression is very variable and differs from person to person. Regular reassessment can be helpful in managing progression and supporting the person with PSP or CBD and their families.

Supporting an individual experiencing cognitive change

It is important to remember that cognitive changes are part of the diagnosis of PSP or CBD and it is not the person choosing to 'be difficult'.

The following general strategies will help in managing cognitive changes:

- Develop structured routines – this will help the person 'know' what is happening and when
- Allow plenty of time for tasks and activities as this will reduce stress
- Try to keep the environment calm with minimal distractions
- Try to remain patient and encourage the person with PSP/CBD to be patient with themselves.

Problems with decision making and processing information:

- For simple decisions try to give choices with just one or two alternatives and try not to use open ended questions. Questions with yes or no answers can be easier
- It can help to remove background distractions (eg TV)
- Break down complex information into smaller chunks and take time to ensure understanding at each stage
- Use pictures or notes if this is easier
- Be prepared to wait for an answer (it can take someone a long time to process information).

Difficulty learning a new task:

- Encourage the person to stop and think
- Break down the task into smaller steps
- Show them what to do by slowly doing the task together.

Help an individual problem solve by:

- Focusing only on relevant issues
- Providing feedback/encouragement
- Doing lots of practice of the steps involved.

Impulsivity:

- Supervise activities, especially walking (people may lack insight into the limitations of their walking ability and the risks associated with their poor mobility); and eating (the use of smaller utensils may help prevent someone 'cramming' food into their mouth). Try to avoid 'Dual-tasking' ie people with PSP/CBD may be distracted from walking if they are doing something else such as talking or holding something, increasing the risks of falls.
- Gently encourage people to stop and think rather than rush an activity
- Recognise that a lack of insight may mean that the individual genuinely does not perceive/believe that the difficulty exists.

Less able to respond appropriately to people/situations:

- Understand that it is likely to be the condition that is causing the inability to respond appropriately. Explain this to family and friends
- The person with PSP/CBD may be less facially expressive than they were and difficulties with eyelid opening may give the false impression that they are asleep
- Recognise that the egocentricity (for example, lack of concern for partner/carer) displayed by the person with PSP/CBD is stressful for the family/carer but it is not personal. Be open to accepting respite care, sitting services, and help from others.

Socially inappropriate behaviour:

This may include, for example, crying or laughing in an inappropriate way and/or making sexually disinhibited comments:

- Explain to family and friends that PSP/CBD can cause such behaviours and that people with these conditions may have reduced understanding of emotion and of social situations
- Respond in a calm manner to anything which is inappropriate
- Consider how to explain the situation to children in an age appropriate way
- Seek advice on whether medication may have a role to play.

Insight

Lack of insight (Anosognosia) is a common feature of PSP and CBD and one that can be especially challenging to understand and manage. It can result in a person taking risks (especially in respect of their mobility), failing to appreciate dangers and lacking empathy and sensitivity to others' feelings.

Lack of insight is quite different from psychological denial; it is not simply denial of a problem but the genuine inability to recognise that the problem exists.

Coping with lack of insight

Coping with a loved one's lack of insight can be very demanding.

It is easier to work around difficulties rather than to try to change a person's behaviour. Strategies often involve changing a situation rather than trying to 'change' (retrain/teach) a loved one.

Tips for carers:

When communicating – be gentle, encouraging and empathetic.

Try to stay calm – focus on the person when voicing concerns and articulate your thoughts in a subtle and positive light.

Remember – lack of insight is not something that your loved one is choosing.

Try not to argue – logical arguments typically will not work. Do not focus on trying to convince the other person to change, rather focus on achieving realistic goals.

Seek help and take care of yourself – reach out to other family members, friends and voluntary or statutory services. Support groups can be helpful. Taking time away will enable you to provide better care and to find the caring role a more satisfying one.

Try to take regular exercise – this releases endorphins that make you feel happy.

Try to stay socially active – recognise that as a carer you might have to be the one who organises things as others may worry about intruding on your time.

Talk to others in a similar situation – caring can be hard work emotionally and physically, contacting others who understand helps reduce isolation.

Learn how to manage stress – maybe try yoga or pilates or relaxation techniques.

Get professional help – do not neglect your own health and raise issues early with your GP and other professionals. They can help arrange services and extra support.

LEAP

A strategy known as LEAP* may be useful, especially at an early stage before lack of insight has become a major issue.

LEAP stands for:

L – Listen to what your loved one is saying, avoid jumping to conclusions and try to get a clear understanding of what they are trying to convey.

E – Empathise with your loved one. This will allow them to open up to you and not feel you are working against them.

A – Agree on things you observe with your loved one about what does/does not work for them. This might involve making changes.

P – Partnership – create a partnership with your loved one to work towards a lifestyle that suits both parties. This means allowing them to be empowered by a partnership based on acceptance. Try to look at the strengths of your loved one and build on these.

**Originally proposed by Dr Amador in 2000 for people with mental illness but adapted, with slight modifications, by Dr Rose who has found it helpful for people with neurological conditions.*

Helping an individual with PSP or CBD discuss their future

Given that cognitive difficulties can be progressive and difficulties with speech and communication increase as the condition progresses it is important to give early consideration to future planning to help ensure the wishes of someone living with PSP or CBD are known and respected. Some people with PSP and CBD may wish to talk about how they would like to be cared for in the later stages of their condition. It is helpful if this is written down in an Advance Care Plan (a statement of wishes). If someone wishes not to have certain treatments or interventions this should be written in an Advance Decision to Refuse Treatment (a legally binding document). Specialist help from those familiar with the condition (eg palliative care staff) should be sought when completing these documents.

If a person wishes to nominate someone else (for example, their partner or a relative) to make decisions about their health and wellbeing thought should be given to having a Lasting Power of Attorney for Health and Welfare in place.

For more information on Advance Care Planning see Information sheet: *Advance Decision guidance notes for people living with PSP and CBD*.

Capacity

Mental capacity refers to the ability to make decisions. If there is doubt as to whether someone has capacity, an assessment should be undertaken. Health and social care professionals can give further guidance on this issue.

As a family it can help to be open and honest about cognitive changes to help ensure everyone has an appreciation of how cognition can be affected by PSP/CBD. Do seek help when needed.

Useful contacts

PSPA Helpline and
Information Service
Tel: 0300 0110 122
helpline@pspassociation.org.uk
www.pspassociation.org.uk

PINNT – a support group for people receiving
artificial nutrition
PO Box 3126, Christchurch, Dorset BH23 2XS
www.pinnt.com
Tel: 020 3004 6193

Further information

We provide other sheets related to this
information: *Advance Decision guidance
notes for people living with PSP and CBD.*

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