

PSP

CREATING A BETTER FUTURE FOR
PEOPLE LIVING WITH PSP & CBD

A GUIDE TO PSP & CBD

FOR GENERAL PRACTITIONERS



INTRODUCTION

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), also known as Corticobasal Syndrome (CBS).

PSPA supports people living with PSP & CBD. The term PSP is used as shorthand for both, unless there are specifics to highlight.

Providing the care and support required for someone living with these conditions can be one of the most challenging experiences faced by a General Practitioner (GP) who may only come across these conditions once or twice in their professional lifetime.

The purpose of this booklet is to provide GPs with information about PSP & CBD with particular focus on:

- Diagnosis and early referral
- Improved quality of life for patients, with effective symptom management aided by increased awareness of the problems/symptoms encountered and treatment options
- Timely involvement of the multidisciplinary team (MDT), including Occupational Therapist (OT), Physiotherapist (PT), Speech and Language Therapist (SALT), Psychology, Cognitive and Palliative care services.

Decisions regarding management of symptoms should be made by the patient, with guidance and support from their neurologist, GP and other members of the MDT.

The goal is to provide an individual, personally tailored approach to care that is coordinated with access to appropriate information and support to help the patient to make informed choices.

This booklet is only a guide. The information is published in good faith but is not a substitute for the advice from the specialist involved in the care of someone with PSP or CBD.

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PRESENTATION – PSP

DESCRIPTION

Progressive Supranuclear Palsy (PSP), was previously known as Steele-Richardson-Olszewski Syndrome. It is a neurodegenerative condition causing both a movement disorder and cognitive problems. It is defined by the accumulation of abnormal tau protein, in neurofibrillary tangles in the brain, resulting in premature death of the neurons. It affects:

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

ONSET

Early symptoms may include:

- Parkinsonism ('extrapyramidal symptoms' of slowness and stiffness but no tremor. Symmetrical, affecting the trunk and neck most)
- Falls – often backwards
- Eye problems with symptoms of 'glare' or fatigue and eye movements up or down are slow and restricted
- Striking facial appearance often with creasing of the forehead (frontalis overactivity) giving a 'startled expression' and reduced blink rate
- Cognitive dysfunction with slowing and mild personality change.

EPIDEMIOLOGY

- Starts above the age of 40 – average onset is 62
- Average time from onset to diagnosis is three years
- Affects men and women. (Slight male predominance).

INCIDENCE AND PREVALENCE

Prevalence: 6.4 people per 100,000 population. This is about 5% of parkinsonian patients seen in a movement disorder clinic.

DIFFERENTIAL DIAGNOSIS

Definitive diagnosis of PSP can only be made by post mortem examination, but specialists can make the diagnosis with 95% accuracy.

The initial symptoms are often mistaken for Parkinson's disease and it may be some time before special symptoms and signs occur that lead one to suspect PSP. But earlier diagnosis is often possible.

The diagnosis is based on history and examination. There are minor mimics, including Parkinson's disease, Multiple System Atrophy, Normal Pressure Hydrocephalus and Vascular Dementia. Tests like MRI, DAT or PET are used mainly as a means to exclude these other conditions that can mimic PSP. The MRI in PSP can show shrinkage of the brainstem (called the 'hummingbird sign' or 'Mickey Mouse' which are specific to PSP) and other areas.

In 2009 different presentations of PSP pathology were described depending on the location of the pathology in the brain and more recently new diagnostic criteria was published. The description given above is characteristic of the most prevalent form of PSP called Richardson's syndrome. A presentation similar to Parkinson's disease where patients can initially respond to dopamine is termed PSP with Parkinsonism (PSP-P).

Other presentations include:

PSP-PAGF – difficulty with gait initiation (PSP with predominant gait freezing).

PSP-CBS – looks like CBS but with a PSP pathology.

PSP-F – frontal cognitive problems (PSP with predominant frontal presentation).

As a consequence, PSP is a very individual condition with a wide range of symptoms and rate of progression. This will require increasing care and support from the multidisciplinary team.

PRESENTATION – CBD

DESCRIPTION

Corticobasal Degeneration (CBD), also known as Corticobasal Syndrome (CBS) is a progressive neurological disorder. CBD is also caused by the accumulation of abnormal tau protein, in neurofibrillary tangles in the brain, but in different parts of the brain, leading to different signs and symptoms from PSP. Many of the treatments for PSP are also helpful in CBD.

The principal areas of the brain affected are:

- The basal ganglia – leading to parkinsonism, dystonia, myoclonus
- The cerebral cortex – affecting vision, perception, coordination and motor skills.

As with PSP, patients with CBD pathology can have various presentations which include those where language is affected (non fluent or agrammatic version of primary progressive aphasia) and where cognition and visual processing is affected (nonfluent variant primary progressive aphasia). Therefore patients need to be assessed and treated individually for the symptoms and difficulties they have.

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 and apraxia (jerky, awkward movements)
- Stiffness and slowness of movement, including dystonia
- Complex unintentional movements of the limb interfering with normal tasks ('alien limb')
- Behavioural changes, apathy being the most common
- Problems with memory, vision and language
- Unlike PSP, CBD is asymmetric – more on one side of the body
- Problems with fluency and effortful speech very common.

EPIDEMIOLOGY

- Starts above the age of 40 – average age of onset 65
- Affects both men and women.

INCIDENCE AND PREVALENCE

- Prevalence: 6 people per 100,000 population.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis is as PSP.

CAUSES AND TREATMENT

CAUSES

Despite advances in the understanding of the biology of PSP the root cause of the disease is still unknown.

Almost all cases appear to be sporadic in a family (i.e. not inherited). Both environmental and genetic influences may be involved to cause PSP. Around 20% of the UK population carry a gene which provides a weak susceptibility to PSP but the triggers for the disease are not known.

Less than 1% of those with PSP have a clearly genetic cause. A variant in the gene for tau protein called the H1 haplotype, located on chromosome 1, has been linked to PSP. This genetic variation is common and is not enough to cause PSP on its own.

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

TREATMENT

There is currently no cure for PSP or CBD but both can be actively treated to reduce symptoms and risks for people with the condition. Management is based around symptom control and quality of life. Hence, early diagnosis and referral to a specialist multidisciplinary team (MDT) is key. Later sections in this booklet discuss medication that can be offered in the management of symptoms.

For drug dosages refer to British National Formulary (BNF) or Palliative Care Formulary (PCF).

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

THE MANAGEMENT OF PSP & CBD

MONITORING AND ASSESSMENT

The MDT offers the best approach to management and improving quality of life. A large number of health and social care professionals will need to be involved with each patient at some stage.

The MDT should use a patient-centred holistic approach to ascertain the patient's physical, social, emotional and spiritual needs.

In order to manage the presenting symptoms and problems the individual may present with, it may be appropriate for the GP to refer to the following members of the MDT:

Parkinson's disease nurse specialist: Medication and management of complex issues

Physiotherapist: Mobility (gait, falls and balance)

Occupational therapist: Mobility, hand function, postural issues and equipment for activities of daily living

Speech and language therapist: Communication aids/cognitive assessment and swallowing difficulties

Dietitian: Weight loss, lack of appetite

Orthoptist: Problems with vision

Continence nurse adviser: Bladder dysfunction

Community matron: Manage complex issues of the condition

District nurse: Support and case management

Community psychiatric nurse: Behavioural problems

Psychologist: Cognition, anxiety, depression and carer support

Specialist palliative care team: Counselling and psychological support. Day therapies, respite, palliation, end of life planning and care

Social worker: Needs assessment, financial review of benefits and local support options

PSPA: Support for individual and family

Information and support can be accessed through PSPA via the helpline on 0300 0110 122 or www.pspassociation.org.uk

MANAGING SYMPTOMS

There are many ways to reduce symptoms. See relevant sections in this booklet.

RED FLAGS – PSP

The common delay in diagnosing PSP & CBD makes it extra important to know the 'red flags' as warning signs for clinical suspicion of PSP or CBD.

Think about PSP & CBD when seeing a patient diagnosed with a movement disorder (e.g. parkinsonism or working diagnosis of Parkinson's disease without tremor). Early falls, a poor response to Levedopa, rapid progression, early changes to personality and cognition.

What to look for in a patient consultation:

1	Falls	Often backwards and without warning
2	Postural instability	Axial rigidity, easily loses balance
3	Slowness of movement	Bradykinesia
4	Motor recklessness	Impulsive, despite problems moving
5	Eye problems	Restricted eye movement, up/down. May find it difficult to walk downstairs with down gaze. Reduced blink, double vision, 'glare'
6	Speech	Slurring of speech, gravelly or lower voice and progressive word finding difficulties
7	Swallowing difficulties	Liquids/and or solids, excessive saliva
8	Cognitive changes	Change in personality, irritability, apathy
9	Emotional lability	Appears to laugh or cry even if not upset
10	No presenting tremor	

If your patient has one or more of these symptoms please refer to Neurology and state PSP in the referral letter.

RED FLAGS – CBD

Remember to think of CBD in patients diagnosed with Parkinson's disease, if the illness and treatment response is not typical. Be suspicious of CBD if the patient shows little response to levodopa or who develops extra features below, or worsens rapidly.

What to look for in a patient consultation:

1	Highly asymmetric 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
8	Poor response to levodopa	Sinemet, Madopar

Unlike PSP, patients with CBD are less likely to experience eye or balance symptoms.

Problems with swallow can occur, usually in late stage disease.

If your patient has one or more of these symptoms please refer to Neurology and state possible CBD in the referral letter.

MOVEMENT AND MOBILITY

The patient will be affected by problems with movement and mobility throughout all stages of the condition including:

- Poor balance
- Early falls and frequent falls, often backwards and without warning
- Unsteady gait with reduced arm swing
- Axial rigidity with marked neck stiffness. This can cause pain and spasm but often surprisingly pain denied by patients despite significant spasm
- Motor recklessness (impulsivity despite the movement disorder)
- Difficulties with complex and fine motor skills
- Bradykinesia (slowness of movement)
- Muscle rigidity
- Loss of ability to maintain weight-bearing position.

CONSIDER

- Assessment to support movement and activities of daily living
- Assessment for a specialist wheelchair
- Assessment for walking aids
- Telecare falls sensors
- Education of carer on likelihood of increased movement impulsivity and decreased safety judgement with disease progression.

MEDICATION TO CONSIDER

- Levodopa (Sinemet, Madopar) or Dopamine agonist (e.g. Rotigotine, Pramipexole, Ripinrole) with faster dose rise than in Parkinson's disease
- Amantadine (caution over 75)
- Botulinum toxin for painful muscle spasm.

REFER ON TO

- Physiotherapist – for individual with PSP or CBD and also carer, for education
- OT.

Whilst impulsive movement seems a mild impairment, individuals exhibiting this require a high level of supervision due to the high risk of falls and constant support from the carer. This may mean that a greater level of support at home is required at this time.

Also see *A Guide to PSP & CBD for Occupational Therapists*.

PAIN

People living with PSP & CBD may have physical pain caused by:

- Muscle rigidity
- Muscle cramps
- Immobility
- Ice-cold sensations in extremities.

MEDICATION TO CONSIDER

- Simple analgesia, e.g. long lasting NSAIDS – paracetamol
- Botox for spasm, dystonia or painful rigidity
- Baclofen or Clonazepam (beware cognitive disturbances, falls risk)
- Amitriptyline (beware cognitive disturbance)
- Gabapentin (neuropathic pain)
- Pregabalin (may also help with sleep).

SKIN SENSITIVITY

- Good skin and pressure care is vital. Refer to the district nurse for pressure-relieving equipment.

REFER ON TO

- Physiotherapist
- OT
- Complementary therapist
- Pain that is hard to manage – specialist pain clinic or specialist palliative care team.

Communication problems may make understanding the type and source of pain challenging and should be taken into consideration.

The PACSLAC is an appropriate assessment tool if communication is difficult.

Involvement of the person's carer and wider family is essential in understanding the nature of the pain and how to alleviate it.

SPEECH AND COMMUNICATION

The majority of people with PSP & CBD will develop communication problems at some point as the condition progresses and changes may happen quickly. Individuals may experience:

- Slurred speech
- Dysarthrophonia – effortful, distorted, loud/quiet speech
- Hypomimia – reduced facial expression – limiting expression of emotion
- Restricted mobility and restricted eye movement may limit communication
- Cognitive deficits – leading to repetitive speech and confusion of 'no' and 'yes' when the opposite is meant
- Echolalia and Palilalia – repeating of stammering for syllables of whole words
- Very erratic speech, with few remaining words intelligible
- Cognitive problems which may lead to withdrawal or difficulty engaging in social interaction, often due to difficulty following pace of conversation
- Problems with executive function may make it easy to be confused
- Slow response times in conversation.

CONSIDER

- Prism glasses available from PSPA which may help with the use of communication aids
- Speech applications on tablets or lightwriters – be aware of cognitive change that may reduce the ability of the person to use them effectively
- Asking important questions in multiple ways to confirm intentions
- Advice to carers about communication strategies.

REFER ON TO

Early referral to speech and language therapist (SALT) who will:

- Arrange for assessment and provision of communication aids and strategies – ask if the SALT can continue ongoing supervision and reduce multiple referrals
- OT – environmental controls.

Liaise with clinical psychologist if cognitive problems are impacting communication.

DYSPHAGIA

Difficulties with swallowing are common in PSP and swallowing changes impact an individual's ability to maintain appropriate nutrition and hydration. These changes can greatly affect the psychological wellbeing of the person living with the condition and their family.

Problems may include:

- Coughing and aspiration when drinking fluids or eating
- Weight loss
- Being at further risk of choking or aspiration if cognitive issues lead to cramming mouth with food and gulping of liquids
- Excess runny or thick saliva
- Potential drooling as a result of poor lip seal and control of facial muscles
- Recurrent chest infections.

Early sensitive discussions regarding PEG feeding should be held with the individual and their family carers to identify what their wishes may be in the future. This discussion is best begun while the individual is relatively fit. The discussion can be revisited later, but beware not to persuade or coerce the individual against their wishes. PEG feeding is for palliation – and can be used alongside oral intake of favorites.

CONSIDER

- Assess nutritional intake and weight
- Provide information on good oral health
- Altered meal patterns can be helpful, i.e. instead of three meals a day take a 'little and often' approach – consult dietitian
- Alter consistency of diet – consult dietitian or SALT
- Possibility of thickening fluids – consult SALT
- Carer support.

MEDICATION TO CONSIDER

Thin runny saliva

- Atropine eye drops administered under the tongue
- Glycopyrronium bromide (glycopyrrolate)
- Hyoscine patches (should be used with caution as are likely to cause confusion)
- Botulinum toxin to the salivary glands.

Thick saliva

- Mucodyne
- Nebulised saline
- Pineapple, apple, papaya or lemon juice
- Check fluid intake.

Dry mouth

- Artificial saliva sprays – Glandosane
- Good oral hygiene – consider referral to specialised dental services.

REFER ON TO

- SALT – swallowing assessment and discussion of PEG insertion
- Dietitian
- OT.

BLADDER AND BOWEL

Problems with the bladder and bowel can range from minimal to severe impairment throughout the condition. Individuals may experience:

- Urinary frequency and symptoms of overactive bladder
- Difficulty initiating flow and poor flow
- Feeling of incomplete emptying of bladder
- Nocturia
- Urinary incontinence
- Constipation
- Diarrhea
- Frequent urinary tract infections.

MEDICATION TO CONSIDER

- Macrogols such as Movicol
- Laxido
- Molaxole or similar as part of bowel management
- Prophylactic antibiotics for chronic UTI's
- Caution against anticholinergics as cognitive and falls risk increased
- Solifenacin or other antimuscarinics.
- Mirabegron for urinary urgency or mild incontinence

REFER ON TO

- Dietitian
- Continence adviser
- OT
- Physiotherapist
- District nurses.

VISION

Problems with vision can include:

- Slower or hypometric eye movement
- Vertical gaze palsy – restricted up or down gaze
- Photophobia glare
- Excessive lacrimation (tearing)
- Blepharospasm
- Decreased blink rate – causing corneal problems and dry eyes
- Eyelid apraxia – inability to open the eyes at will, may lift brows up to 'help' the eyelids.

CONSIDER

- Education of family and carer regarding possibility of falls due to problems with downward gaze
- Prism glasses (available from PSPA)
- Ptosis props and Lundie loops to help alleviate blepharospasm and apraxia of eyelid opening
- Tinted wraparound sunglasses.

MEDICATION TO CONSIDER

- Artificial tears or eye sprays
- Ocular lubricants, especially at night
- Botulinum toxin for blepharospasm and apraxia of lid opening.

REFER ON TO

- Orthoptist
- OT.

COGNITION

Changes in executive function, behaviour and mood are common and include:

- Slowness of thought processes
- Impaired ability to manipulate acquired knowledge
- Difficulty concentrating
- Poor judgment and planning
- Apathy/disinterest/aggression
- Lack of motivation
- Impulsivity (including recklessness when they move)
- Poor sleep
- Impaired recognition of emotions and lack of empathy
- Changes in mood, including depression
- Emotional lability and personality changes.

Timely assessment is key and a base line assessment should be carried out.

REFER ON TO

- Cognitive disorders clinic
- Clinical psychologist
- Community nurse
- OT.

Supporting carers is vital. Care givers often feel unprepared and understandably struggle. Access to psychological support for both the individual and the carer/family should be offered as appropriate. Education for the carer and family may be useful in managing challenging or unexpected behaviours.

The various cognitive effects of PSP can lead to the reduction or loss of mental capacity. It is important for people living with PSP to be given the opportunity to make advance decision whilst they still are able to do this.

You may also wish to refer to *A Guide to Cognition for Health and Social Care Professionals*.

SPECIALIST PALLIATIVE CARE

PSP & CBD are rapidly progressive conditions with a wide range of symptoms. A palliative care approach – good communication with patient and family/caregivers, shared decision making and goal setting and symptom management – is essential from diagnosis. Access to early specialist palliative care is helpful in allowing families to plan for the future, discuss wishes and make choices that are right for them. The specialist palliative team are able to provide symptom management and psychosocial support and may be appropriate from early on in the disease progression. Sensitive and careful discussion is important as many people associate palliative care/hospice with end of life and the benefits of earlier involvement should be discussed and explained. Inclusion of the specialist palliative care team early on helps to support both those affected by the conditions and the professionals involved.

It is important that all people with PSP & CBD are placed on the GP Supportive Care Register and discussed regularly in a multidisciplinary Gold Standards Framework meeting. This will ensure that planning future care can be undertaken whilst linking the opinions of the numerous professionals involved.

Discussions about Advance Care Planning/Advance Decision to Refuse Treatment ensures that patient's wishes are respected at each stage of the disease. The progressive nature of the cognitive changes seen in both conditions often impact on an individual's ability to maintain capacity as defined by the Mental Health Capacity Act 2005.

Areas that may need to be discussed and considered are:

- Where a person wishes to be cared for
- Alternative methods of feeding e.g. feeding tubes
- Whether they would want to have cardiopulmonary resuscitation
- Antibiotic therapy.

Hospices often run a day service which allows patients to access therapies such as massage, develop a rapport and gain input from palliative care specialists. This will help ease the transition to advanced disease and end of life, as well as provide significant support and respite for carers. Speaking to the local hospice about what can be

offered to PSP & CBD patients is fruitful for the patient/carer and you as their GP.

When end of life care is required the person living with the condition and their families will be better supported by staff they already have a rapport with, and who understand the complexities of a neurodegenerative illness.

THINKING AHEAD – ADVANCE CARE PLANNING (ACP)

The GP is in an ideal position to help the patient talk through options for their care preferences at the end of life stage. Early assessment for inclusion on the Palliative Care Register and access to specialist palliative care is needed due to the rapidly degenerative nature of both conditions.

Early discussions regarding ACP are particularly important because of communication difficulties that people can experience. This makes it vital to discuss and seek their views on treatment and management early, when they are still easily able to express and communicate their desires and wishes.

ADVANCE DECISION TO REFUSE TREATMENT (ADRT)

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

Before making an ADRT the contents should be discussed with the GP to ensure that the patient is clear regarding which treatments they wish to decline and they have a full understanding.

If this is made, it is good practice for a copy to be kept by the GP surgery with the patients record.

It is important that all people with PSP & CBD are placed on the GP Supportive Care Register within the practice.

EMERGENCY HEALTHCARE PLAN (EHCP)

EHCP's are helpful as a source of information and guidance for GP's and emergency services. Ideally made by MDT and/or GP in collaboration with the patient and their families.

An EHCP can be invaluable to lay out 'what to do' guides in each scenario when out of hours GP's or ambulance teams are contacted. This can help prevent unwanted admissions

END OF LIFE

Many people living with PSP or CBD and their families fear end of life. However the most common cause of death is from aspiration pneumonia, which is typically peaceful and pain free if managed well – 'slipping away' without choking or distress. Towards the end of life anxiolytics (such as benzodiazepines, lorazepam orally/sublingually or midazolam buccally or by injection) and analgesia should be considered in line with good end of life care principles according to local protocols and palliative care departments.

Family and carers will need practical and emotional support. Care plans and information must be shared by all members of the care team and adequate nursing cover maintained.

The end of life stage can be difficult to predict. This stage may last a few days or several weeks. Indicators may include:

- Reduced levels of consciousness
- Inability to eat and drink
- Severe infection that could not be treated in the home environment (hospital admissions may have previously been refused)
- Fall or major fracture
- Rapid and significant weight loss.

Being a carer for someone with a long-term condition can be overwhelming. There can be many strains upon the relationship which can be particularly challenging for carers, especially if the person with the condition experiences changes in personality, or a loss of apathy or ability to recognise emotions in others.

It may be appropriate or helpful to arrange some time to talk to the carer without the patient present, perhaps, whilst blood tests are being done. This can help offer support, find out which issues are burdening or distressing for them (these may be different than the patient's concerns) and allow time to provide support networks.

Carers need advice, support and information to help them to make sense of what is happening, to enable them to continue in their caring role, and help them plan for the future.

It is important to check with the carer at every stage that they feel able to continue with their caring role.

Carer stress is common and they may experience any of the following:

- Guilt
- Disbelief
- Helplessness
- Anger
- Depression
- Sleep disturbances
- Inability to concentrate
- Weight loss/gain
- Inability to cope
- Financial pressures
- Loss of social networks
- Anxiety.

MANAGEMENT

- Support carers by regular and separate assessment of their own needs
- Allow time for the carer to discuss how they are feeling
- Refer on to social services for a Carers Assessment – this should anticipate need and must be outcome-based and reviewed on a regular basis
- Ensure access to community matron or district nurse
- Carers should be registered as carers on the practice register and be given priority for treatment
- Respite – discuss if required and facilitate this if necessary
- Signpost to PSPA and carers associations for support.

Many carers will not ask for help until they reach crisis point and GPs are in a position to prevent this.

People with PSP & CBD should be booked for double appointments due to slowness of speech and movement. Home visits will be necessary as the disease progresses.

People with PSP & CBD, their families and carers often experience considerable psychological and emotional distress. Loss of hope, fear, lack of control, anxiety and frustration are just some of the emotions that people may experience. All of these will affect the patient's other holistic needs.

Ensure those affected by PSP & CBD have an appropriate level of information about their condition. Be guided by the patient and carer's preferences for how much information they would like.

Distress is likely to vary throughout the course of the disease and patients should be referred for counselling, if available. Identify an individual to provide ongoing support and information. This may include support from local psychological services or the local hospice.

People may lose their social support network due to changes in occupation, hobbies, relationships and in their physical abilities. It is common for people to become socially isolated, which can have an impact on their physical and psychological wellbeing.

- Consider referring to hospice day therapy services
- Provide information on support available from PSPA.

Depression is not always easy to differentiate from sadness, but treatment is likely to have a positive effect. Consider screening for anxiety and depression in patients and especially carers where persistent low mood or hopelessness is expressed and treat according to local guidelines for mental health conditions: remember the carer may also be a patient in their own right.

RESEARCH

PSPA funds and promotes ground-breaking research to improve the lives of people affected by PSP/CBD and is dedicated to supporting the search for treatments.

PSPA Research Strategy 2020 – 2025 comprises of four elements:

1. Prioritise PSPA research.
2. Support the research stars of the future.
3. Engage patients and carers in research.
4. Enhance communication, collaboration and change.

More information on PSPA Research Strategy 2020 – 2025 can be found at: <https://pspassociation.org.uk/research/research-strategy>

A major step towards the achievement of our aims is the UK PSP Research Network.

The PSP Research Network is made up of seven centres of expertise – UCL, Oxford, Cambridge, Newcastle, Manchester, Southampton and Newport.

The network will allow pooling of samples and data. It will also provide a basis for communication and collaboration (across different area of biomedical research).

These centres are now involved in a long-term study called PROSPECT. This study involves the collection of serial biological samples and clinical information from a cohort of patients over several years, enabling researchers to make new discoveries based on the way that PSP changes over time. A separate cross-sectional arm of the study will collect one-off blood samples, generating a resource for investigating indicators of disease in blood as well as genetic data.

For further information on PSPA research projects visit www.pspassociation.org.uk/research

BRAIN DONATION

There are a number of brain banks across the UK collecting brain tissue from people with neurodegenerative diseases such as PSP & CBD.

Brain tissue is one of the most important resources for neuroscience research and provides invaluable insights for researchers into the causes of PSP & CBD and how the disease progresses. Hopefully, in the long-term, this will enable researchers to also develop a much-needed diagnostic test and an effective treatment.

Information on other local brain banks can be found at www.hta.gov.uk/guidance-public/brain-donation

FURTHER INFORMATION

For health and social care professionals from PSPA

An Introduction to PSP & CBD

An information leaflet for the newly diagnosed explaining both conditions and services available through PSPA.

A Professionals Guide to PSP & 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. 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 need. <https://hscpguide.com/>

A Professionals Guide to PSP – A Professionals Guide to CBD

These guides include an overview of standards of care and best practice for all those working with, providing services for and supporting people with PSP & CBD.

A Guide to Cognition for the Primary Healthcare Team

This booklet contains information on cognitive and behavioural change, as well as practical tips on management.

A Guide to PSP & CBD for Physiotherapists

This guide provides physiotherapists with information about PSP & CBD and to guide and inform their practice and intervention.

A Guide to PSP & CBD for Occupational Therapists (OTs)

This booklet aims to provide OTs with information about PSP & CBD and aims to address issues at different stages of progression. It also provides practical suggestions to aid therapeutic intervention.

Red Flag Posters for PSP & CBD

Posters to share with colleagues to help identify the early signs of PSP & CBD.

ACKNOWLEDGEMENTS

Professor James Rowe, Consultant Neurologist, Cambridge University Centre for Parkinson-plus and Addenbrookes Hospital, Cambridge

Dr Boyd Ghosh, Consultant Neurologist at University Hospital Southampton NHS Foundation Trust and Salisbury NHS Foundation Trust. Honorary Teacher at the University of Southampton

Dr Sarah Agyekum, GP Retainer, London

Dr David Oliver, Consultant Physician in Palliative Care – Medway Community Healthcare

Anna Kent, Neurological Conditions Clinical Specialist, Neurological Conditions Clinical Specialist Team (NCST), Central & North West London NHS Foundation Trust

Dr Louise Wiblin, Senior Neurology Consultant, South Tees NHS Trust

Dr Alistair Church, Associate Specialist, Royal Gwent Hospital.

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

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WE WELCOME YOUR VIEWS

PSPA encourages feedback about any aspect of the information we produce. If you'd like to help us by reviewing future versions of this or other resources, please email us on helpline@pspassociation.org.uk



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Registered in England
number: 2920581

Registered charity numbers:
England and Wales 1037087/
Scotland SC041199

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