

Progressive
supranuclear Palsy

PSP

**A Guide to
PSP and CBD
for
Occupational Therapists**

Working for a world free of PSP

A GUIDE TO PSP AND CBD FOR OCCUPATIONAL THERAPISTS

INDEX

	Page
Introduction	4
The PSP Association	5
About PSP and CBD	6
Multi-Disciplinary Management of PSP	8
Guidance for Occupational Therapists	9
Impairments	11
Occupational Therapy Intervention:	12
➤ Diagnosis and early stage	12
➤ Mid stage	14
• Transfers	14
• Mobility	17
• Stairs	18
• Falls	18
• Feeding	19
• Personal Care	20
• Communication	21
• Vision	21
• Behavioural and Cognitive Change	22
• Carer Support	23
➤ Palliative stage	24
Case Study	26
Conclusion	27
Useful resources	28
References	29

INTRODUCTION

This booklet forms the first of a new series of publications for health and social care professionals, to help them support people who have Progressive Supranuclear Palsy (PSP) or Cortico Basal Degeneration (CBD). (The PSP Association supports both conditions and unless otherwise specified, references to PSP include CBD throughout this guide).

The booklet aims to provide Occupational Therapists with information about PSP and CBD and to describe how the common impairments may affect typical activities and occupations. Through expert opinion and by drawing on literature from comparable progressive neurological conditions, the guide aims to address issues at different stages of the disease process and to prove a practical, useful aid to therapy.

This *Guide for Occupational Therapists* was written by Madeleine Quine (PSP Development Officer for Scotland and trained Occupational Therapist), Joanne Hurford and Kate Morton (Occupational Therapists at the National Hospital for Neurology and Neurosurgery, London). The authors have direct experience of working with people with PSP and CBD and have drawn on these to develop the guide.

We are most grateful for the assistance of many families with PSP and CBD who have contributed, and for the additional professional input provided by:

- *Professor Andrew Lees, Consultant Neurologist at the National Hospital for Neurology and Neurosurgery, London, Director of the Reta Lila Weston Institution of Neurological Studies, University College London, and Chairman of the Progressive Supranuclear Palsy Association's Medical Advisory Panel.*
- *The PSP Nurse Specialists of the PSP Association*
- *The Therapy Services Department, National Hospital for Neurology, UCLH NHS Trust*
- *Fiona Lindop, Derby PCT*
- *Kate Roberts, Nottingham PCT*
- *Millie Ebanks, UHCW NHS Trust*

This Guide will be reviewed annually and we would welcome your feedback for improvement, which should be sent to:

Madeleine Quine, Development Officer (Scotland), The PSP Association
E-mail: madeleine.quine@pspeur.org Tel: 0794 088 8627 or c/o the address below.

© The PSP Association, December 2008
The PSP Association, PSP House, 167 Watling Street West,
Towcester, Northants NN12 6BX
Tel: 01327 322 410 Email: psp@pspeur.org Website: www.pspeur.org

Working for a world free of PSP
Registered charity 1037087

THE PSP ASSOCIATION

The Mission of The PSP Association is to conquer Progressive Supranuclear Palsy (PSP) and an associated condition, Cortico Basal Degeneration (CBD). It works in the interests of people living with these conditions, now and in the future, to:

- Promote and sponsor **research** worldwide.
- **Support** affected families across Europe.
- Provide information and build **awareness of PSP and CBD** among the health and welfare services, and the general public.

Support for people with PSP/CBD and carers from the Association includes:

- A telephone support service, linking to a team of PSP Nurse Specialists for information and advice.
- A comprehensive information pack.
- Local support groups and networks across the UK and Ireland.
- The PSP Association Magazine, three times a year.
- An annual PSP Symposium held at different locations in the UK.
- The PSP Website and Forum.

Support for health and social care professionals includes:

- Access to the PSP Nurse Specialist telephone support service.
- Training and PSP awareness sessions arranged on request.
- Access to the other services outlined above.

For access to the PSP Nurse Specialist Telephone Helpline, to arrange a training or awareness session or to obtain a membership form, see inside last page.

ABOUT PSP AND CBD

What are PSP and CBD?

Progressive Supranuclear Palsy (PSP), and the closely related but rarer Cortico-Basal Degeneration (CBD),² are brain diseases involving progressive nerve cell loss, particularly in the basal ganglia and brain stem that control many motor functions. The loss is linked to an over-production of the Tau protein, leading to the formation of neurofibrillary tangles that progressively distort or block motor signals from the brain to various parts of the body.

How many people in the UK have PSP?

The prevalence demonstrates there are in the region of 4,000 people diagnosed with PSP³ in the UK, yet the incidence and duration of illness means there are over 10,000 people with the disease. Consequently, there are over 6,000 people mis or undiagnosed with PSP. Likewise, this means PSP is more common than the better known Motor Neurone Disease.

What are the main symptoms?

Symptoms vary widely between individuals, but PSP progressively affects balance, movement, vision, speech and swallowing.

Many people also suffer behavioural changes that can range through apathy, depression, irritability, laughing or crying for no reason, or the inability to handle complex issues. However, it is important to realise that the intellect is largely intact, unlike in dementia.

Unexpected backward falls and the inability to look down are common symptoms of PSP that are unusual in other conditions.

Other early symptoms may include stiffness or rigidity in the muscles in the back of the neck, depression, apathy, dislike of bright lights, tunnel vision, tiny cramped handwriting, fear of falls and loss of judgement.

The rate of progress of symptoms varies widely from person to person, but is often quite rapid. A routine and regular assessment of needs is essential. This is especially important after an event that has resulted in hospitalization such as a damaging fall, when a sudden decline in capability can be quite significant.

What is the typical life expectancy?

Average life expectancy from the onset of symptoms is some five to seven years, but there is considerable variation and some patients progress much faster.

What treatment is available?

There is currently no effective treatment to slow or halt the progression of PSP. Treatment is palliative, aimed at maximizing the available quality of life for the person with PSP and their direct carer.

²For the purposes of this leaflet, PSP and CBD are referred to as 'PSP'

³Based on a prevalence of 6.4 per 100,000. Information provided by The PSP Association

ABOUT PSP AND CBD

How is PSP diagnosed?

A clinical diagnosis is made after observing the range of symptoms to form an overall picture. It is important that suspected cases are referred to a Neurologist. Post-mortem examination confirms that the clinical diagnosis is well over 90% accurate.

PSP can affect people as young as 40 though usually affects people who are over 60. Although research is encouraging, there are no clear diagnostic markers and an accurate diagnosis in the early stages can be difficult. In the early stages, PSP and CBD symptoms can resemble those of more common conditions such as Parkinson's, Alzheimer's or a stroke.

Can PSP be inherited?

About a quarter of the UK population has the genetic predisposition to PSP, but very few develop the disease. There are probably many other trigger factors, and these are being actively re-searched.

There is no proven case of a child in the UK inheriting PSP from a parent and the probability of inheriting PSP is extremely low.

MULTI-DISCIPLINARY MANAGEMENT OF PSP

Due to the complex and multifactorial nature of PSP, individuals and their carers should be offered an integrated assessment and planning of their health and social care needs to make informed decisions about their care.⁴ Support from a multidisciplinary (MDT) team with a variety of health care specialists is essential in providing this integrated assessment and will be required throughout the disease progress. Good, consistent communication between the health care professions and family is crucial in the provision of good quality care, and addressing the individual's and any carers' needs.⁵

Key members of the team providing support from diagnosis to the palliative phase include the General Practitioner, Neurologist and Clinical Nurse Specialist. Other key team members include the Occupational Therapist, Physiotherapist, Speech Therapist, Social Worker and Community Matron. A key worker system to trigger referrals and aid communication will help to ensure the timely involvement of the appropriate professionals as the disease progresses.

The diagram identifies some of the frequently involved multidisciplinary team members in the individual's care and emphasises the need for good communication.

Even if the need for individual specialist input is negligible at the point of diagnosis, the unpredictable and possibly rapid progression of symptoms means that early contact between specialists and patients is essential.

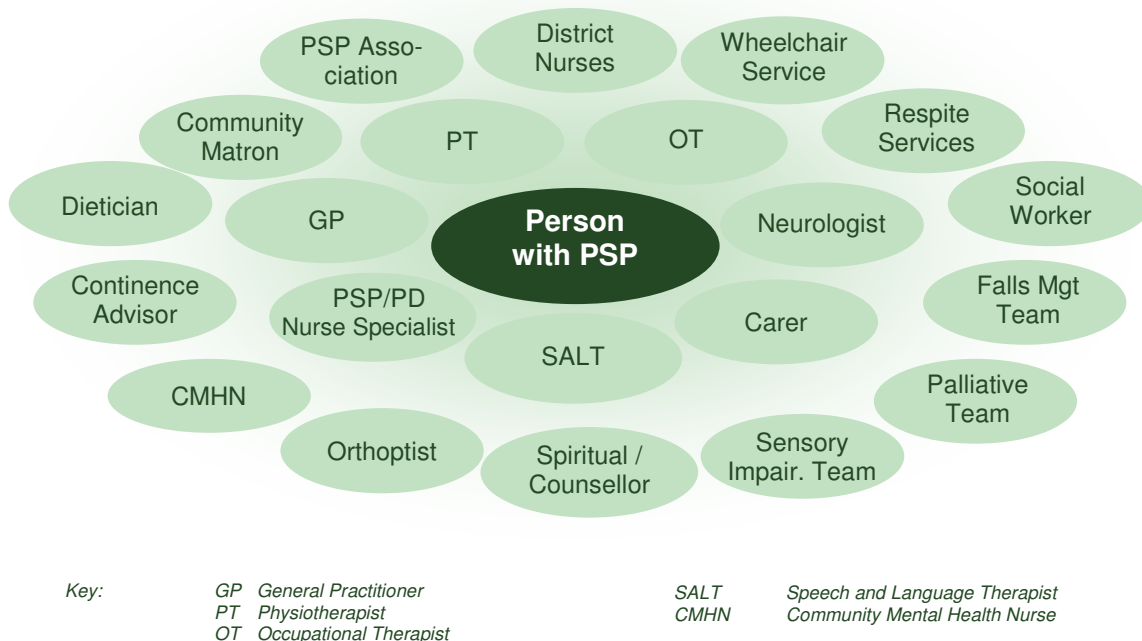


Fig 1: The Multi-disciplinary Team for PSP and CBD (Quine, Hurford and Morton)

⁴Department of Health, 2005

⁵Astbury and Rose 2005, NICE 2006

GUIDANCE FOR OCCUPATIONAL THERAPISTS

Introduction to Occupational Therapy with PSP and CBD

This booklet aims to provide OTs with information about Progressive Supranuclear Palsy and Cortico Basal Degeneration, and demonstrate how important occupational therapy is as an intervention. The guide will describe how the common impairments may affect typical activities and occupations, and will address practical issues at different stages of the disease process.

The current evidence available in the field of PSP primarily focuses on medical management and diagnosis. There is no primary research evidence to support OT intervention with PSP. Even Parkinson's disease (PD) which is similar and more widely recognised has limited research evidence available to support OT intervention. However, it is recognised that OT is an important part of the integrated care team that the patient will need to access in order to maintain independence and quality of life.⁶

OT Models and Treatment Approaches

Although PSP is a complex neurological condition, therapists should feel confident in drawing on their core skills to assess and understand the impact of impairments on function; utilising problem solving and clinical reasoning skills to provide effective intervention.⁷ A client centred model such as the Canadian Model of Occupational Performance can be a useful tool in understanding the individual's key issues in a holistic manner.⁸

Motor learning theory and a task-orientated approach have been advocated in the therapeutic management of PD.⁹ These support a functional approach to treatment in which movement patterns and components of tasks are practiced in relation to functional activities. It may be beneficial to consider using these approaches in the early stages of PSP to minimise impairment, develop effective task specific strategies and adapt functional goal-orientated strategies to changing task and environmental conditions.¹⁰

Quality of life in individuals with PSP is often affected due to the progressive, disabling nature of the disease and decreased life expectancy. OT intervention should focus on goals that support the individual, carers and family to try to minimise the impact of the disease, thereby adding to their quality of life.¹¹ Treatment decisions should be made on a case-by-case basis considering the individual's cognitive status, capacity to learn, stage of disease, degree of impairment, the difficulty of the task, the environment and the individual's changing needs and goals.¹²

A Delphi study of OT for individuals with PD (Deane et al 2003) identified four main roles for OTs: problem solver, educator, net-worker and supporter. Further to this, Jain et al (2005) identified a framework for intervention for progressive neurological conditions (see fig 1).

⁶Dean et al, 2003

⁷Law et al, 2002

⁸CAOT 1997 cited by law et al, 2002

⁹Trail, 2008

¹⁰Shumway-Cook and Woollacott, 2001

¹¹Schrag et al, 1994

¹²Trail, 2008

GUIDANCE FOR OCCUPATIONAL THERAPISTS

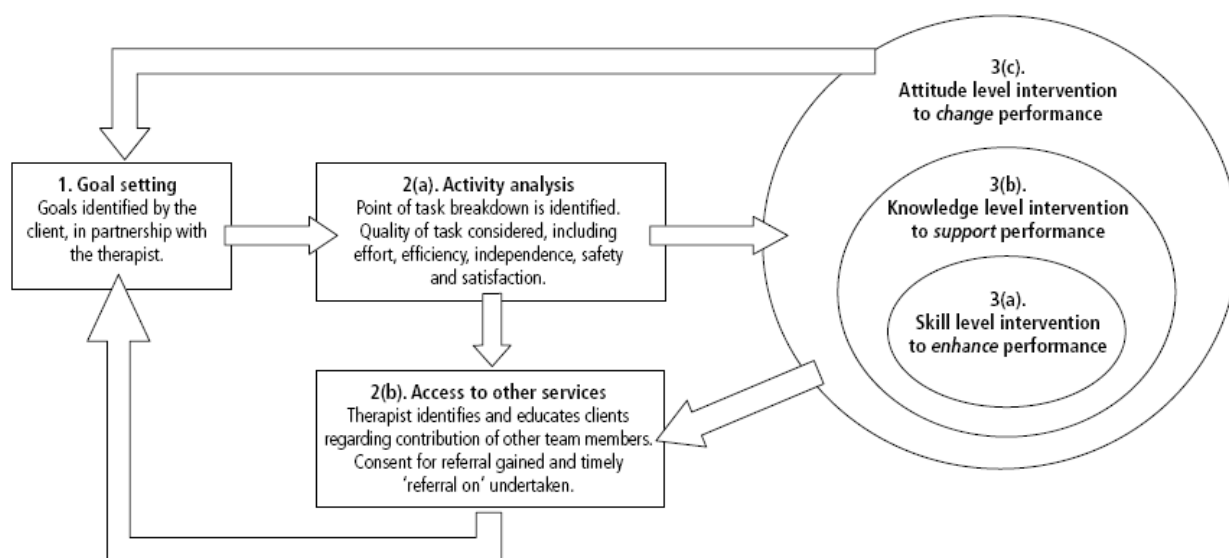


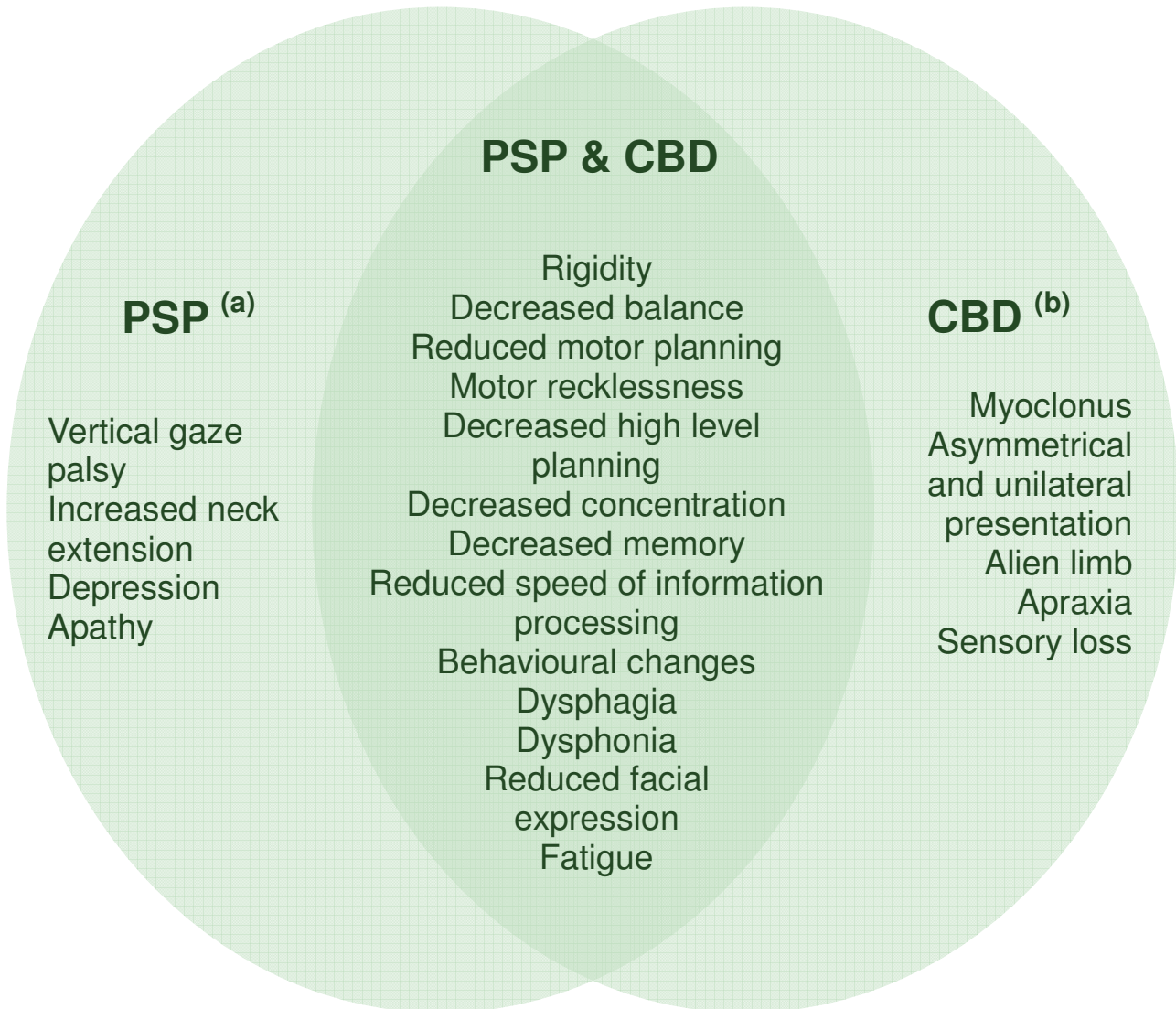
Fig 2: Framework for intervention Jain et al (2005) Reprinted with permission from BAOT

Considerations for assessment and intervention

- Gather as much background information prior to commencing assessment as possible, to strengthen your initial assessment.
- Consider the key impairments and activity and participation difficulties and explore this within the initial interview.
- Use a theoretical OT model to guide and underpin treatment intervention.
- Acknowledge and address the carer and family's needs within the assessment process.
- Be mindful of the rate of disease progression and how this will influence your intervention.
- Allow sufficient time for the individual to discuss their difficulties, taking into account their stamina levels and cognitive processing skills. Be sensitive to their needs, returning to complete the assessment later if required.
- Consider the individual's insight into their current strengths and weaknesses and how this may affect their safety.
- Consider the individual's attitude to their diagnosis and their acceptance of aids and services.
- Establish a list of main concerns and prioritise treatment goals.
- Discuss functional ability and observe performance within every day tasks.
- Utilise The PSP Association's telephone service to liaise with PSP/CBD Nurse Specialists for further advice and support for the individual and their carer.
- Be aware of local support services available to the individual and carer and make early referrals to them.
- Arrange regular reviews of the individual's functional ability due to the fast progression of the disease. Carers are unlikely to call for support before they have reached crisis point, which then makes intervention more difficult.

IMPAIRMENTS

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



Reference: (a) Burn and Lees 2002
(b) Welling et al 1998

Fig 3: Common impairments in PSP & CBD

Symptoms become increasingly severe as the diseases progress, and an early referral to the local palliative care team should be initiated early on in the disease process even though their direct support may not immediately be required

OCCUPATIONAL THERAPY INTERVENTION DIAGNOSIS AND EARLY STAGE

As with most chronic progressive neurological conditions, people's reactions to a new diagnosis can vary significantly. The individual may experience a variety of emotions ranging from fear of the future, anger, denial and hopelessness through to acceptance.¹³ Due to the complex nature of the condition, a definitive diagnosis is often delayed.¹⁴ Symptoms may already be affecting the individual's performance and a degree of acceptance and adjustment may have already begun.

In considering Jain et al's (2005) framework for OT intervention, the OT role as an educator, net-worker, problem solver and supporter are discussed for this client group:

Educator:

The Condition: Therapists working with patients with PSP do not need to be experts in the area to provide basic advice on the condition. However, with information widely available on the internet, individuals will vary from having very little to vast amounts of information about their diagnosis. It is therefore important for the therapist to have a basic understanding of the types of symptoms that are likely to be experienced through the progression of the disease and the time frames in which these may occur. In understanding this, it will enable the therapist to judge where the individual is considered to be within this process and therefore plan intervention accordingly. However, whilst it may be useful to plan ahead for the individuals' future needs, this can often be difficult for the individual or family to accept and you should be sensitive in how this is approached.

Resources and Information: The newly diagnosed individual often lacks knowledge of health, social care and charitable resources available to them. At this stage, individuals should be directed to The PSP Association who are a key resource in providing valuable and practical advice and support to the individuals and their carers. A key role of the OT at this stage is to provide information on community services and resources that may help them throughout the disease process.¹⁵ As well as the support provided by the Local Authority, such resources include benefits, community transport, Blue Badge Scheme, the Disabled Living Foundation and equipment suppliers.

Driving The newly diagnosed individual with PSP should contact the Driver and Vehicle Licensing Agency (DVLA) and their insurance company to inform them of their diagnosis as soon as it is confirmed because impairments may affect their safety to drive. This is a legal requirement.¹⁶

The individual should be reassured that reporting the diagnosis to the DVLA does not mean that their driving licence will be withdrawn. The DVLA will conduct an assessment from the information provided by the individual and medical team. They may request attendance at a driving assessment centre before making a decision.

Networker:

Referrals to Other Agencies: Therapists working with individuals with suspected or newly diagnosed PSP may be based in the acute hospital setting or the community. It is important that the individual's longer-term needs are considered and the appropriate referrals are made early within the disease process to community services such as the Community Rehabilitation Team, Social Services or local wheelchair services.¹⁷ Agencies should be made aware that referrals are urgent because the condition is likely to progress rapidly.

¹³Playford, 2003

¹⁴Scaravilli et al, 2005

¹⁵NICE, 2006

¹⁶Your Personal Guide to PSP, Leaflet "Getting About", The PSP Association

¹⁷Department of Health, 2005

OCCUPATIONAL THERAPY INTERVENTION DIAGNOSIS AND EARLY STAGE

Problem Solver and Supporter:

Activities of Daily Living (ADLs): In the early stages of the disease process individuals are likely to be independent and able to sustain most activities of daily living; but these will often be more effortful. Therefore, advice on fatigue management strategies to assist them in planning and prioritising their ADLs is useful. Complex or fine motor tasks such as handwriting may become difficult and it is important to address these within OT assessment. Functional assessments within specific tasks are required in order to trial and identify the most suitable strategies or aids at this stage.¹⁸ Keeping recommendations simple is often most effective.

Work: PSP affects people as young as 40 years therefore support in the work place and managing their working role may be a priority or concern for the individual. Key areas that the occupational therapist may assist with include:

- Support and advice on continuing to work if this is important to the individual and is a practical objective. The individual does not necessarily have to give up work immediately just because they have PSP.
- Support regarding the disclosure of their diagnosis to others and their employers, if this is a concern.
- Exploring difficulties in the work place to identify reasonable adjustments to the environment or role that optimises their performance. These may include technological adaptations, assistance with travel, introduction of frequent breaks and support within the work place.
- Signpost-to-work resources such as Access to Work or Job Centre Plus.
- Advising the individual on their rights and responsibilities under the Disability Discrimination Act (2005).
- Maintaining a balanced view of what the client would benefit from as well as the employer's need to maintain a productive service.
- Reviewing the individual's satisfaction with their work-life balance.
- Advising on application for benefits if this is appropriate and signposting to Job Centre Plus for support.

¹⁸Fisher, 1999

OCCUPATIONAL THERAPY INTERVENTION MID STAGE

People with PSP are often at this stage before a definitive diagnosis is confirmed. This is often when a referral to Occupational Therapy will be made as activities of daily living become increasingly difficult. Although individuals may not be able to carry out all the elements of a task, it is important to maximise participation to maintain a sense of self worth and competence.¹⁹ Likely areas of difficulty include:

Transfers

People with PSP struggle with transfers due to limb and trunk rigidity, lack of cognitive planning for the task, vertical gaze palsy and increased neck extension.²⁰ Sit to stand is especially difficult from low surfaces and they will often drop back into the chair when sitting (sit 'en bloc') making them at risk of hitting their head if a chair is against the wall. When standing up; the 'rocket sign' is evident as they race off in an unsteady manner, putting them at risk of falls.²¹

No specific equipment has been identified for this client group and the clinical reasoning to identify appropriate strategies and equipment will need to consider the person, environment and task.²²

Education to the individual and the carer on how to carry out the transfers safely, focusing on slowing down, aligning self correctly and executing the transfer using normal movement patterns, is an essential component of the treatment approach.²³ However, due to impairments with judgement and reasoning, patients may not retain techniques practiced or information about how to use equipment in the first instance, so repetition is likely to be required. It is therefore important for the therapist to consider this when making recommendations, as equipment may not always be the most appropriate option to optimise function. Also, equipment needs may change frequently due to the progressive nature of the disease.

¹⁹Graff et al, 2006

²⁰Morris et al, 1999

²¹Burn and Lees, 2002

²²Law et al, 2002

²³Parkinson's Disease Society, 2007

OCCUPATIONAL THERAPY INTERVENTION

MID STAGE

Top Tips for Transfers

Chair Transfers

- Raise the chair to an appropriate height: aim for at least 90 degrees at the knee.
- Ensure the chair is stable and that armrests are at a suitable height/position to enable the patient to push up (remember individuals with CBD may only be pushing up with one arm in the initial stages).
- Educate the patient and carer on sit to stand techniques, emphasising controlled normal movement and even weight distribution to facilitate standing.
- If the individual still struggles with sit to stand consider the use of a rise/recline chair.

Bed Transfers

- The use of a bed leaver is beneficial to aid rolling and rising in bed.
- A satin half sheet placed at the mid third of the bed can also be useful with turning and rolling in bed as it reduces friction. However, careful consideration should be given as this may contribute to the person's risk of falling during transfers.²⁴
- Educate the patient and carer on bed transfer: breaking down the transfer into rolling, dropping legs off bed and pushing up into sitting.²⁵
- Consider the height of the bed and location within the room.

Toilet Transfers

- Be aware of the impact of motor recklessness (individuals appear to be unaware of their poor balance, and mobilise quickly and unsafely) and rigidity when assessing the appropriate toilet equipment.
- Freestanding toilet equipment will probably be inappropriate due to risk of falls.
- Consider how continence issues such as increased frequency, night time toileting, and decreased speed of accessing the toilet can be managed.

Bath Transfers

- Assess the safety of appropriate bath equipment, taking into account the potential impact of rigidity, wandering limbs, reduced visual field, backward falls and motor recklessness.
- Bath boards would probably be inappropriate due to risk of falling backwards.
- Trial use of a swivel bather or any electronic bath aid with a supportive seat, however these may only be a short-term solution due to the rapid progression of the disease.
- Consider the need for early referral for major adaptations such as a level access shower to enable continued bathing as other options may soon become unsafe.

²⁴Parkinson's Disease Society, 2007

²⁵Parkinson's Disease Society, 2007

OCCUPATIONAL THERAPY INTERVENTION MID STAGE

Car Transfers

- Educate the patient and carer on positioning the car to allow the door to be opened fully and to transfer onto an even surface of appropriate height.
- Educate the patient and carer on car transfer techniques i.e. getting legs square against the seat prior to sitting down.
- A firm cushion may be used to raise the height of the seat or a wedge to level off a sloping seat.
- A clip-on handle to the car door to aid transfer or a flexible fabric turning-disc with a non-slip base may be useful aids to trial, to assist with moving legs in and out of the car.
- If car transfers become increasingly difficult and unsafe individuals may wish to travel in a wheelchair taxi or consider adapted cars with support from their regional driving assessment centre.

OCCUPATIONAL THERAPY INTERVENTION

MID STAGE

Mobility

PSP damages the regions of the brain associated with balance control, often leading to slowness in movement, motor recklessness - particularly in sitting and standing up, muscle rigidity, and deterioration in posture, gait and stamina.²⁶ However, despite the rapidly progressive nature of the condition, rehabilitation and education may help to maintain balance functions and slow the decline in mobility.²⁷

Individuals with CBD present with unilateral and asymmetrical onset of impairments that have different functional implications for mobility than PSP. The loss of use of one hand will impact on ability to use a walking frame. As CBD progresses, it is no longer asymmetrical and mobility will need to be reviewed regularly. In common with PSP, there may be a disturbance of eye movements, although it is less striking than with PSP.

Physiotherapists are expert at assessing mobility and suitability of walking aids. It is important that OTs liaise closely with their physiotherapy colleagues to discuss the functional implications of mobility within the individual's different environments and tasks in order to optimise safety.

Problems with mobility can be compounded by a motor recklessness, which means individuals may appear to be unaware of their poor balance, and mobilise quickly and unsafely.²⁸ If the individual also suffers with vertical gaze palsy, a common problem with PSP, then they will be unable to look down and scan for hazards as they mobilise. Education around the set up of the home environment is essential for the individual and the carer to aid safe mobility. This should include:

- Adequate space for walking aids and turning circles
- Clear pathways and minimisation of trip hazards such as loose wires and rugs
- The padding or removal of sharp edges and hard surfaces in likely fall areas, as it is unlikely that falls can be prevented totally.

As the disease progresses, difficulties experienced with mobility may become stressful for carers. A wheelchair may be a beneficial option but the decision can be seen as a loss of mobility and an indication of the progression of the disease. The OT plays a key role in assessing and referring for the wheelchair and appropriate accessories such as head supports, pressure cushions and arm rests/lap tray for alien limb management. The OT will also need to provide psychological support to the individual and carer with the acceptance of this new equipment, discussing the benefits to both of them. As the individual becomes more dependent and a wheelchair is required for indoor mobility, access in and around the house will need to be reviewed.

²⁶Burn and Lees, 2002

²⁷Steffen et al, 2007; Suteerawattananon et al, 2002

²⁸Burn and Lees, 2002

OCCUPATIONAL THERAPY INTERVENTION

MID STAGE

Top Tips for Walking Aids:

- Consider a neurological physiotherapy referral for a period of rehabilitation and education.
- Weighted walking frames have been suggested as a useful mobility aid for people with PSP. An example is the U step (see resources list for details), a reverse braking mobility aid using the concept of shifting weight forwards to reduce the risk of falling backwards when holding onto the frame. However, there is currently no primary evidence to support the effectiveness of weighted walking frames and their provision will need to be considered on an individual basis. These are not yet routinely available through statutory services and a special order or funding may need to be obtained.
- Built up/wedge shoes that help shift the weight forwards with the aim of reducing falls backwards have been suggested by some people with PSP. However, there is limited primary evidence to support the use of wedge shoes and if being trialled the therapist should consider the patients mobility and safety at times when they may not be wearing the shoes such as night time toileting.
- A one handed walking aid such as a stick or tripod may be of benefit for individuals with CBD in the early stages.
- In view of cognitive dysfunction, wall fixed rails around the home environment may in some cases provide a more effective solution for mobility problems.
- Assessment within the home environment is the key to the identification of appropriate solutions for people with PSP.

Stairs

The combination of backwards falls and difficulty with downward eye gaze make stairs a particular hazard. Assessment of safety on stairs will need to be completed and recommendations made accordingly, for example, a second stair rail, major adaptations (stair lift or through floor lift), bed being brought down stairs or support of a carer to supervise stair mobility. Liaison with physiotherapy colleagues for stairs assessment is recommended.

Falls

Falls backwards are a significant problem and individuals who experience earlier falls are likely to have a faster progression in the severity of the disease. Falls are often sudden and without warning, and individuals with PSP are at higher risk of skull and rib fractures, unlike PD where hip fractures are most common due to the direction and rigidity during the fall.²⁹

Falls are a major consequence of the disease and become a constant part of the individual and carers' lives. As such, it should be a priority for the OT to address. (See NICE guidelines for falls management, 2004, for further information.)

²⁹Williams et al, 2006

OCCUPATIONAL THERAPY INTERVENTION

MID STAGE

Top Tips for Falls Management:

- Early referral to falls management group.
- Referral and liaison with Physiotherapy colleagues.
- Education on getting up from the floor for individual and carer.
- Advice on equipment available such as pendant alarms and other integrated technologies such as sensor mat and carer voice alert systems.
- Education to the individual and carer to concentrate on one task at a time to reduce the risk of distraction when mobilising and transferring.
- Protective garments such as helmets, hip protectors and rib protectors may be useful for some individuals who are frequently falling, however there is currently no evidence to support their effectiveness at preventing fractures. If considering these a referral to the local Orthotist may be beneficial.

Feeding

The characteristics of severe vertical gaze palsy and cervical dystonic neck posturing into extension or neutral erect position are distinguishing symptoms of PSP. The former affects the individual's ability to see and locate food on the plate in order to load the spoon/fork and bring it to the mouth, and the latter increases aspiration during swallow.³⁰ Environmental aids are likely to be required to bring the plate within the individual's visual field and joint assessments with Speech and Language Therapy colleagues are essential to determine the qualities of the individual's swallow.

Top Tips for Feeding:

- Be aware of the individual's swallow status before carrying out any assessment.
- A variable height table or raising the height of the plate with a phone book or similar may be useful to locate the plate within the individual's visual field.
- Plate guards provide additional sensory input when loading the fork/spoon.
- Insulated bowls/plates may be useful for individuals who require longer to feed themselves.
- For individuals with mild alien limb syndrome, trial limiting the degree of freedom and the effects of gravity on the arm by modifying the environment i.e. type of cup, propping arm on table, table height and seat height.
- For individuals experiencing more severe alien limb syndrome consider one-handed strategies/equipment such as rocker knife and use of dycem mats.
- The OT has a significant role in educating formal and informal carers on the set up required for maximum independence with feeding. This may be within the home, hospital or care home environment.
- Work closely with the Speech and Language Therapist and Physiotherapist to assess the impact of the individual's seated posture and how this may affect the quality of their swallow. More supportive seating may be beneficial.
- Consider a referral to a dietician for advice on energy and fluid intake if this is a concern.
- Consider the effects of the environment as potential distractions may impact on the individual's performance.

³⁰Leopold & Kagel, 1997

OCCUPATIONAL THERAPY INTERVENTION

MID STAGE

Personal Care

Assessment of personal care will be required, taking into account the main difficulties experienced such as motor recklessness, vertical gaze palsy and processing difficulties in PSP, and apraxia, myoclonus, involuntary movement and alien limb in CBD.³¹ Assessment and intervention should be ongoing, in order to grade the level of assistance needed. A client centred approach should be maintained as the retention of a degree of independence in a component of a task can add to self-esteem and quality of life.

Top Tips for Personal Care

- Angled mirrors in the bathroom may aid the individual's ability to search and locate items needed and to self monitor grooming tasks such as shaving and cleaning teeth.³²
- Easy access to personal care items and the provision of appropriate seating will help to minimise the risk of falls.
- Velcro fastenings or elastic waistbands may make dressing and undressing easier.
- Consider how involuntary movement dyspraxia may impact on safety and use of tools within tasks.³³
- Keep surfaces and work environments as clear and free from distraction as possible.
- Consider alternative toiletry items to reduce the complexity of task such as pump action toothpaste and soap, or electric toothbrushes and razors.
- Dressing aids for lower half may be of benefit, such as sock aids and long handled shoehorns.
- Providing advice and assistance to carers on how to supervise, prompt and assist the individual. This might include :
 - Reducing visual and auditory environmental distraction
 - Breaking down the task into simple steps
 - Providing short simple verbal prompts
 - Allowing extra time for responses to prompts and the completion of each stage
- Early Social Services referrals for care support may help to ease the carer's strain with personal care tasks.
- Consider referral to a District Nurse or Continence Advisor if continence issues are affecting the individual or carer's quality of life.

³¹Nabir and Pharm, 2003

³²Gentry, 2003

³³Graham et al, 1999

OCCUPATIONAL THERAPY INTERVENTION

MID STAGE

Communication

Individuals with PSP are likely to have problems with communication. This can be due to:

- Decreased volume of speech
- Dysarthria
- Fixed facial expression
- Inability to gain eye contact due to vertical gaze palsy
- Decreased speed of information processing
- Difficulty with drooling.

These symptoms can give the inaccurate impression that the individual is disinterested in conversation. It is important that everyone involved with the patient understands this. Communication issues also include difficulty with handwriting, typing, the use of the telephone and e-mail.

Top Tips for Communication:

- Liaise with the Speech and Language Therapist about communication strategies and potential aids. The OT's role may involve how to maximise movement to best use communication aids. Due to gaze palsy and cognitive deficits AAC (Alternative and Augmentative Communication) equipment may not be suitable.
- Educate carers on the impact of impairments on communication.
- Allow extra time for responses to questions.
- Reduce distractions during assessment.
- Split sessions into manageable time lengths to avoid the impact of fatigue on communication.
- Provide cognitive handwriting strategies to increase written letter/word size, if this is relevant to the individual.
- Encourage individuals to sit down when using the telephone to reduce risk of falls.

Vision

Vertical gaze palsy is a key clinical sign of PSP. Other impairments may include:

- Blurred or double vision (diplopia)
- Tearing
- Reduced blink rate
- Photosensitivity
- Interrupted smooth tracking and steady gaze
- Reduced ability for the eyes to converge while watching objects move closer.

Assessment by a Neuro Ophthalmologist and Neuro Optometrist is very important and this should be requested by the individual's medical team. The use of Fresnel prisms on glasses to shift gaze downwards may be recommended by the ophthalmologist. Double vision can be treated by patching one eye or putting tape over one lens. Tinted glasses can be useful for photosensitivity.

The OT's role should focus on practical tips to manage visual impairments:

OCCUPATIONAL THERAPY INTERVENTION MID STAGE

Top Tips for Visual Impairments:

- Advise on the benefit of good lighting within the home environment.
- Ensure walkways and work surfaces are clutter free.
- Improve visual colour contrast within the home.
- Flat colours are easier to see, so changes to wall paper/carpets may be advisable.
- Downward tilted mirrors in areas such as the bathroom and kitchen may be useful to counteract vertical gaze palsy when carrying out tasks.
- Use of bookstands that hold reading material at eye level may be useful.
- Registering as blind or partially sighted may be appropriate.
- Referral to local Sensory Impairment Team may be beneficial if vision is significantly affecting functional ability.
- Provide advice on resources such as talking books or newspapers.

Behavioural and Cognitive Changes

Psychological changes in PSP can be an early symptom and a misdiagnosis of dementia or psychotic illness can be common.³⁴ Cognitive impairment is commonly characterised by frontal lobe dysfunction in PSP that differs from patterns found in PD, Alzheimer's disease and Huntington's disease. Primarily the individual's intellect will remain intact. The difficulties listed below may lead to conflict with family members and carers and can significantly increase the carer's strain on a day-to-day basis.

Chiu (1995) identifies cognitive and behavioural changes in four categories:

Cognitive impairment	Forgetfulness Slowing of thought processes Impaired ability to manipulate acquired knowledge Perseveration Decreased flexibility of conceptual and motor sequencing
Affective and behavioural changes	Irritability Impulsivity Emotionally labile Apathy
Psychotic symptoms	Paranoid thoughts Hallucinations are occasionally present
Sleep disturbance	Insomnia

Top Tips for Behavioural and Cognitive Changes:

- Educate the individual and carer to simplify day-to-day activity by reducing visual and auditory stimuli, and breaking down the tasks into smaller, more manageable steps.
- Educate the carer to allow time for the individual to respond to prompting.
- The use of memory and orientation aids may support both the individual and the carer.
- Referral to day care centres, respite services and the community mental health team can be an integral part of holistic management.

³⁴Chiu, 1995

OCCUPATIONAL THERAPY INTERVENTION MID STAGE

Carer Support

PSP will have a major impact on the whole family, not just the individual. A progressive disease changes the present and future life of the spouse or children particularly those who are still living at home as they are likely to be required to alter their behaviour and take on new roles.³⁵ Carers responsible for assisting individuals with dementia type symptoms often experience helplessness, social isolation and loss of autonomy.³⁶

A recent OT study focusing on individuals with mild to moderate dementia identified the benefits of a goal-directed rehabilitation programme that included carer training. This showed a significant improvement in the carer's sense of competence and the improved daily function of the individual.³⁷ The importance of the carer's role should not be underestimated and should remain an integral role of all assessments.

The carer requires education on the condition and resources available to them to enable them to feel as in control of their situation as possible.³⁸

The PSP Association. Encourage the individual or carer to become a member of the PSP Association (see pages 4-5).

Technology. Telecare can enable carers to avoid direct 24-hour supervision and to know that they will be informed if an emergency occurs. Telecare systems comprise a range of sensors positioned throughout the home to alert the carer, or a monitoring centre that will organise an agreed response. Telecare systems may be purchased privately or provided by social services, community nurses, or housing officers from district councils and housing associations.

Top Tips for Supporting Carers:

- Initiate early referrals to social services to complete a carer's assessment.
- Discuss with the MDT early referral to the palliative care team to provide carer support.
- Provide information on non-statutory support organisations such as Crossroads who may be able to provide additional time out and support for the carer.
- Consider day care services if appropriate.
- Ensure the carer has the practical skills and appropriate equipment required to carry out the task in a safe manner that does not put them at risk. Referrals to a community rehabilitation team or social services OT may be beneficial if extra support is required in this area.
- Ensure the individual and carer has adequate advice and information on their entitlement to financial benefits and exemptions.
- Ensure communication between the carer and all team members is consistent and regular using a key worker system if appropriate.

³⁵Playford, 2003

³⁶Graff et al, 2006

³⁷Graff et al, 2006

³⁸NICE, 2004

OCCUPATIONAL THERAPY INTERVENTION PALLIATIVE STAGE

PSP is a progressive illness with a limited life expectancy of approximately 5-7 years from onset of symptoms³⁹ and the individual will become more reliant on their carer as the disease progresses. A referral to the local palliative care team should have been initiated early on in the disease process.⁴⁰ The palliative care team will be able to support the psychological and spiritual concerns of a progressive condition as well as providing support with symptom control, arranging respite/hospice care if needed and assisting the family to prepare for bereavement.⁴¹ There may be an OT linked with the palliative care team who will be a good resource for managing functional ability at end of life.

Probable difficulties towards end of life⁴²:

- Increasing immobility leading to increased stiffness, pain on movement and possible contractures
- Susceptibility to pressure sores
- Communication may be extremely difficult, adding to the individual's frustration. However they often remain aware and alert of what is going on around them.
- Double incontinence and urinary tract infections are common
- Cough reflex weakens, therefore chest infections and aspiration pneumonia become more likely
- Increasing immobility leading to increased stiffness, pain on movement and possible contractures.

OT input should focus on the safe management of decreasing functional ability:

- Supportive seating system such as tilt-in-space wheelchair
- Equipment to assist with safe transfers such as a hoist
- Support with appropriate positioning in bed
- Pressure care needs
- Ensure personal care needs are effectively met

Collaborative working with the multi-disciplinary team (MDT) is vital at this time to ensure the individual and carers are supported with a consistent approach to meet their changing needs. A continuing care assessment and social services assessment are likely to be required at this time to ensure appropriate support and funding is in place.

³⁹Astbury & Rose 2005

⁴⁰NICE, 2004

⁴¹Astbury & Rose 2005

⁴²Astbury & Rose 2005

OCCUPATIONAL THERAPY INTERVENTION PALLIATIVE STAGE

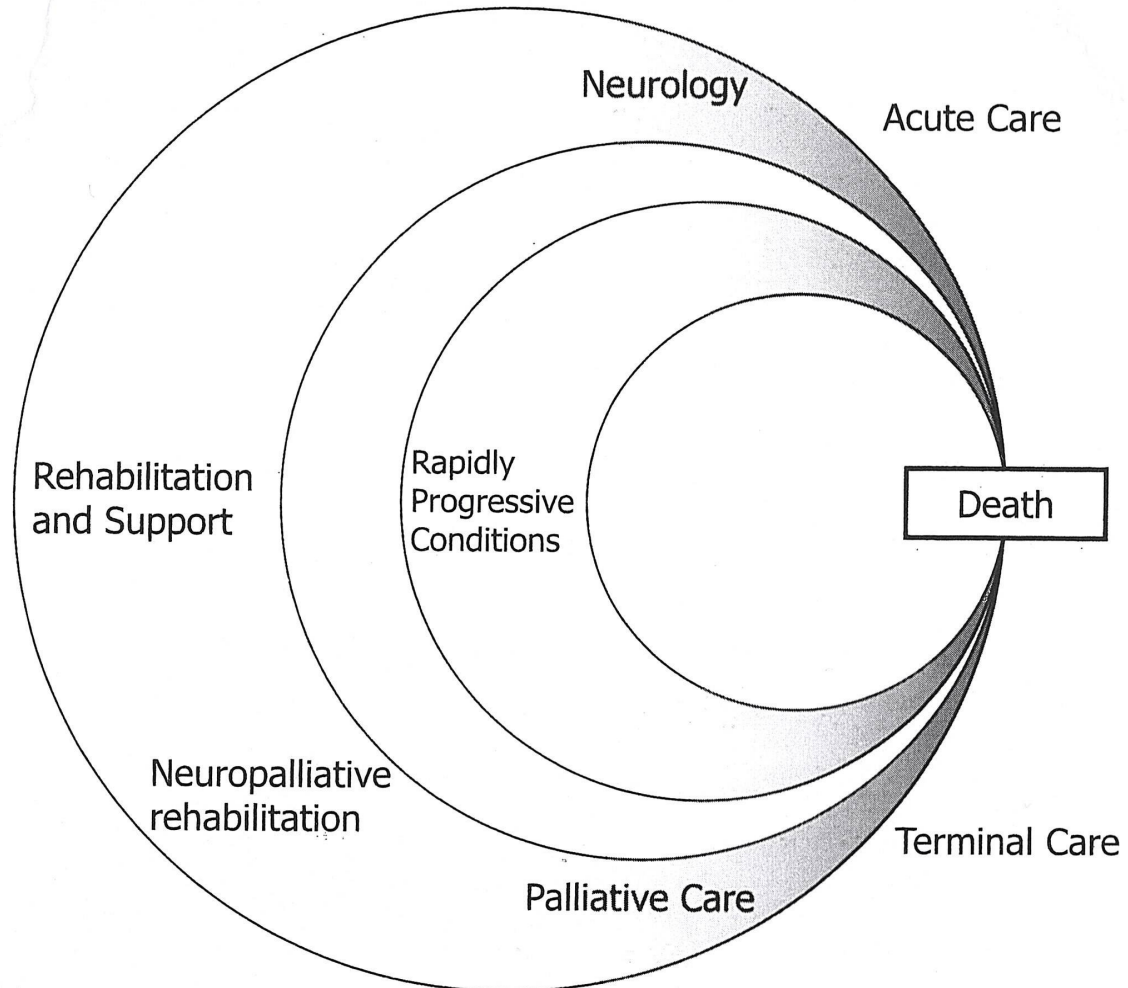


Fig. 4: Model of service interaction for Neurology, Rehabilitation and Palliative Care Services (National Council for Palliative Care, October 2007; permission sought)

CASE STUDY

Jack is a 65 year old man with a 7-year history of difficulties with his speech, including stammering, dysarthria and reduced volume. He was recently diagnosed with PSP during an in-patient admission for investigations and medication review. During his admission, he was referred to OT for assessment of falls management. He has previously been assessed by a Social Services OT.

History of Condition:

A year after his initial problems Jack started experiencing generalised stiffness and falls backwards. After 4 years, his visual field had greatly reduced. During the last year, there has been a rapid deterioration in his functional ability.

Social Situation:

Jack lives with his wife in a privately owned bungalow. His wife and daughter are very supportive. Jack is able to mobilise indoors, but experiences frequent falls. These restrict him from accessing the community. He would like to be able to participate in social activities with his family with increased confidence. Jack's wife reports that she is experiencing increased stress as a carer and feels that she may benefit from more emotional support.

Impairments:

- Reduced balance
- Vertical gaze palsy
- Increased rigidity and stiffness
- Motor recklessness
- Dysarthria
- Dysphonia
- Stammering
- Reduced volume of speech

Activity and Participation Difficulties:

- Difficulty turning in bed; currently uses bed leaver
- Difficulty standing from low chairs; chair already raised at home
- Frequent falls, up to 3 times a day
- Mobilises outdoors with stick plus supervision
- Requires floor fixed toilet frame for independent toilet transfers
- Requires supervision for showering
- Difficulty with dressing lower half, especially socks
- Dependent on wife for domestic tasks
- Misses food on near side of plate
- Requires extra time for communication of basic needs.

CASE STUDY

OT Intervention:

Jack remained an in-patient for 5 days and during this time the OT saw him for three sessions. Areas addressed during these sessions included:

- Providing information on The PSP Association and other services available to provide support to Jack and his wife
- Advice on bed transfer, trialling use of satin sheet technique and socks with grips
- Advice on feeding: trialling use of plate guard on near side of plate to aid putting food on fork
- Advice on dressing: trialling use of sock aid
- Assessment with kettle tipper and perching stool as Jack wished to make own hot drinks, but had previous falls carrying out this task
- Advice on falls management: getting up from floor, use of mobile phone to call for help
- Liaison with Physiotherapist and Speech and Language Therapist
- Referral to local wheelchair services for wheelchair to aid outdoor mobility and increase ability to access social events with family
- Referral to local community Neuro OT and Physiotherapist to practice transfers and falls management techniques within own environment.

CONCLUSION

Individuals with PSP and CBD contend with an array of rapidly changing impairments affecting their ability to engage in daily activities within the home and the community. Each individual requires a holistic assessment by an OT to address how these impairments are affecting them and their carers at the different stages of the disease process. OTs can use their clinical reasoning and learnt knowledge of the condition to treat this client group effectively even if they have limited experience in this area.

This booklet has aimed to give an understanding of the impairments associated with PSP and CBD and how they impact on function, along with practical tips for OTs to use in the treatment of their clients. At present, there are no studies that support specific OT intervention with this client group and there is a need to promote research to examine and test the effectiveness of some of the interventions described. The evidence for this booklet has been taken from expert opinion and evidence related to comparable progressive neurological conditions. The PSP Association is always available to discuss any further issues that have not been addressed within this booklet.

USEFUL RESOURCES

The PSP Association PSP House 167 Watling Street West, Towcester, Northants NN12 6BX
Tel 01327 322410 or <http://www.pspeur.org/>

AbilityNet 0800 269 545 AbilityNet is a registered charity that helps disabled people access their computers and the internet by using adaptations and tailor-made services.
Access to Work scheme gives you and your employer advice and financial support with extra costs which may arise because of your needs.
www.direct.gov.uk/en/DisabledPeople/Employmentsupport

Alzheimer's Disease Society <http://www.alzheimers.org.uk/site/index.php>
Carers UK Equal Partners has produced a briefing – Spotlight on telecare – which covers the pros and cons of the technology, how carers can get telecare support, and what needs to happen to make this technology more widely available. The briefing is available to download from www.carersuk.org/ or call **0161 953 4233**.

Crossroads 0845 450 0350 Crossroads is a service offering a trained Care Support Worker to take over from the Carer to give them a break.

Cure PSP Society for Progressive Supranuclear Palsy in America <http://www.psp.org/>
Department for work and pensions The Department provides benefits and services <http://www.dwp.gov.uk/lifeevent/benefits>

Listening Books 020 7407 9417 Abridged and unabridged books on tape for people with disabilities

Respite care 0845 644 4932 www.bettercaring.co.uk Better caring is dedicated to helping care professionals and consumers find care homes in the UK.

RNIB Talking Book Service Unabridged books on tape or CD. Tel **0845 762 6843** or www.rnib.org.uk/talkingbooks

Talking Newspaper Association: National newspapers and magazines in electronic format. Tel **01435 866 102** or www.tnauk.org.uk

The Disabled Holiday Directory 01348 875592 A holiday organization that provides a wide range of locations and accommodation styles for people needing accessible holidays.

The Disabled Living Foundation provide free, impartial advice about all types of disability equipment and mobility products for older and disabled people, their carers and families.
www.dlf.org.uk or call **0845 130 9177**.

The Telecare Services Association has a website designed to help service users, their carers and family to learn more about telecare and its providers. Call **01634 846 209**.

U step weighted walking frame, available from www.attainability.co.uk

Remap Charity Custom made equipment for disabled people <http://www.remap.org.uk/>
or Tel: **0845 130 0456**.

REFERENCES

- Astbury T, Rose M. (2005) Chp 12 Caring for a person with Progressive Supranuclear Palsy in Swinn (2005) *Parkinson's Disease Theory and Practice for Nurses* ed Whurr Publishers London.
- Burns D, Lees A. (2002) Progressive Supranuclear Palsy: Where are we now? *Lancet (Neurology)*, Vol 1 Oct, 359-369.
- Chiu H. (1995) Psychiatric Aspects of Progressive Supranuclear Palsy. [Review] *General Hospital for Psychiatry*, 17(2): 135-43.
- Cure PSP Society for Progressive Supranuclear Palsy, PSP Disease Management, Section four, *A guide to living with PSP*. Cure PSP. Available from http://www.psp.org/materials/disease_management.pdf (accessed on 10.07.08).
- Deane HO, Ellis-Hill C, Dekker K et al. (2003) A survey of current OT practice for Parkinson's disease in the UK. *British Journal of Occupational Therapy*. 66(5):193-200.
- Department of Health DOH (2005) The National Service Framework for Long-term Conditions available from http://www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAndGuidance/DH_4105361 accessed on 10.08.08.
- Disability Discrimination Act (2005) Available from http://www.opsi.gov.uk/acts/acts2005/ukpga_20050013_en_1 accessed on 12.09.08.
- Fisher AG. (1999) *Assessment of Motor and Processing Skills*. Ft Collins,CO: Three Star Press.
- Gentry T. (2003) Adapt-Ability with PSP: An Occupational Therapist's perspective. *PSP Advocate*, Second quarter. Available at http://www.psp.org/pdfs/2nd_issue_2003.pdf [accessed on 10.07.08].
- Graham N, Zeman A, Young A, Patterson K, Hodges J. (1999) Dyspractia in a patient with corticobasal degeneration: The role of visual and tactile input to action. *Journal of Neurology and Neurosurgery and Psychiatry* 67: 334-344.
- Graff M, Vernooij-Dassen M, Thisjssen M, Dekker J, Hoefnagels W, Olde Rikkert M. (2006). Community based occupational therapy for patients with dementia and their caregivers: randomised control trail. *British Medical Journal*. Vol 333 (7580) pg 1196-1201.
- Jain S, Kings J, Playford, D. (2005) Occupational therapy for people with progressive Neurological Disorders: Unpacking the black box. *British Journal of Occupational Therapy* 68(3).
- Law M, Polatajiko H, Baptiste S, Townsend E. (2002) Core Concepts of Occupational Therapy. In: *Enabling Occupation, An Occupational Therapy Perspective*. CAOT publications ACE Chapter 3:29-56. Canadian Association of Occupational Therapists, 2nd Ed. Ottawa, Canada.
- Leopold N, Kagel M. (1997) Dysphagia in Progressive Supranuclear Palsy: Radiologic Features, *Dysphagia* 12: 140-143.
- Morris H R, Wood N W, Lees A J (1999) Progressive Supranuclear Palsy (Steele Richardson Olszewski disease) *Post graduate Medical Journal* 888: 579-584.
- Nabir Mir G, Pharm M. 2003 Cortico Basal Ganglionic Degeneration *JK Practitioner*. 10 (\$) 253-255.
- NICE National Institute for Health and Clinical Excellence. (2004) Clinical practice guidelines for assessment and prevention of falls in older people. Available from <http://www.nice.org.uk/nicemedia/pdf/CG021fullguideline.pdf> at 10.09.08
- NICE National Institute for Health and Clinical Excellence (2006) Parkinson's Disease: Diagnosis and management in Primary and Secondary Care. NICE Clinical Guidelines 35. Available from <http://www.nice.org.uk/nicemedia/pdf/cg035niceguideline.pdf> at 24.08.08

REFERENCES

- Parkinson's Disease Society (2007) *The Occupational Therapist's guide to Parkinson's disease in The Professional Guide to Parkinson's disease*. Parkinson's Disease Society UK.
- Playford D. (2003) *Neurological Rehabilitation in Parkinson's Disease*. Martin Dunitz. London.
- PSP Association (2007) *Your Personal Guide to Progressive Supranuclear Palsy*. PSP Association Northants, UK.
- Schrag A, Selai C, Quinn N. (1994) Health-related Quality of Life in patients with Progressive Supranuclear Palsy. *Annals of Neurology Vol 35 (2):237-9*.
- Shumway-Cook A, Woollacott MH. (2001) *Motor control*. 2nd ed. Philadelphia: Lippincott Williams & Wilkins.
- Steffen TM, Boeve BF, Mollinger-Reimann LA, Peterson, CM. (2007) *Long term locomotor training for gait and balance in a patient with mixed Supranuclear Palsy and Corticobasal Degeneration*. Vol 87 980, 1078-1087.
- Suteerawattananon, M. MacNeill, B. Protas, EJ. 2002. Supportive treadmill training for gait and balance in a patient with Progressive Supranuclear Palsy. *Physical Therapy*. Vol 82 (5), 485-95.
- Trail M. (2008) *An Occupational Therapy Model of Treatment for Parkinson's disease* in Trail M, Protas E and Lai E (2008) *Neurorehabilitation in Parkinson's disease: an evidence based treatment model*. Slack Enc. New Jersey, USA.
- Welling GK, Litvan I, Jankovic J, Granata R, Mangone CA, McKee A, Poewe W, Jellinger K, Chaudhuri K, D'Olhaberriague L and Pearce R: (1998) Natural history and survival of 14 patients with Corticobasal Degeneration confirmed at post mortem examination. *Journal of neurology, neurosurgery, psychiatry* 64, 184-189.
- Williams D, Watts H, Lees A. (2006) Predictors of falls and fractures in bradykinetic rigid syndromes a retrospective study. *Journal of Neurology, Neurosurgery and psychiatry* Vol 77 468-479.

The PSP Association

PSP House
167 Watling Street West
Towcester, Northants NN12 6BX

Telephone:
01327 322410

Nurse Specialists:
Kat Haines (North West & Ireland) 01995 601533
Jill Lyons (South West & Wales) 01934 842366
Samantha Pavey (South East) 01747 841461
Katie Rigg (North East & Scotland) 01434 382564

Fax:
01327 322412

Email:
psp@pspeur.org

Website:

The PSP Associations Registered Office: PSP House,
167 Watling Street West, Towcester, Northants, NN12 6BX
Registered No: 2920581

Registered charity Numbers: England & Wales 1037087 / Scotland SCO 41199