Pathway of Care for PSP
A guide for Health and Social Care Professionals
Introduction

In 2011, the PSP Association commissioned Neurological Commissioning Support to develop a Care Pathway for PSP. This guide arose from that work and now describes acceptable and deliverable standards of care and best practice guidelines for the management of PSP by all disciplines. Contributions to the guide were made by people living with PSP, consultants and clinicians with expertise in PSP and CBD.

The guide comprises the following sections:

- A set of Standards of Care: before and at diagnosis, during disease progression, and at end of life
- Health and social care guidelines for best practice at four stages of disease progression
- Symptom ‘snapshots’ of different symptom aspects, depicting likely prevalence and suggested interventions
- A visual care pathway depicting the range of services and support required by a person with PSP.

In response to this work, in April 2012 the PSPA launched its new strategy “PSPA: SUPPORT FOR FAMILIES”.

Our vision is to make life better for people with PSP and CBD. In the next three years we will create closer relationships with people with PSP and CBD and we will deliver realistic, practical help and advice to ensure that they receive all the services and benefits they should. The PSPA will now focus on acting as advocates for individual families.

In so doing, it will enhance the capabilities of those individual health and social care professionals who are dealing with PSP and CBD in the present, rather than attempting to educate a large population of those who might be required to do so at some time in the future.

People living with PSP experience symptoms at different points, in different ways and to varying severities from person to person, and this can make acceptance and adjustment very difficult for the individual and their family. It can also make planning and providing services across both health and social care hugely challenging.

CBD is a related progressive neurological disorder characterised by nerve cell loss or deterioration and atrophy of multiple areas of the brain. Although some characteristics differ from PSP, and overall progression is generally slower, symptom management has much in common with that recommended for PSP, particularly the need for coordinated care, access to information and early palliative support.

We hope this guide will help those supporting people with PSP and CBD to plan coordinated and timely care management, and enable greater access to services for people affected by the conditions. The PSP Association supports people living with both conditions and recommends that until a specific pathway for CBD is developed, this care pathway should inform best practice for CBD also.

The PSP Association
Progressive Supranuclear Palsy (PSP) is a progressive neurological disease caused by the death of nerve cells in the brain. It causes severe and unpredictable impairments which have an enormous impact on the individual and their family. Average life expectancy is six to seven years from symptom onset for the majority of people.

As PSP progresses, it causes difficulty with balance, movement, vision, speech, swallowing and cognition. The individual’s ability to walk, talk, feed themselves or communicate effectively with the world around them decreases, thought processes often slow down, yet people with PSP usually remain mentally aware.

PSP is a very individual disease, with symptoms being experienced at different points, in different ways and to varying severities from person to person. This can make acceptance and adjustment very difficult for the individual and their family, whilst requiring increasing levels of care which often falls predominantly to family members, particularly spouses.

People with PSP often find that their communication and cognitive difficulties mean that social inclusion is a challenge. General lack of understanding of both the condition and the needs of those living with it, means that access to appropriate advice, support and information is a constant struggle. The planning and provision of services is further complicated by the variation of symptoms and the severity of the condition from person to person.

### Standards of Care

The PSP Association has set out the following Standards of Care as a guide, to ensure people with PSP receive the best possible care, support and advice from the outset.

#### Before diagnosis

- People with suspected PSP should be referred quickly to a movement disorder specialist or consultant specialising in Parkinson’s.
- PSP is frequently misdiagnosed, often as Parkinsonism. Suspicion of a misdiagnosis due to presentation of PSP symptoms requires prompt referral for further assessment by a consultant neurologist.
- Initial symptoms which may indicate PSP include:
  - Problems with balance, unsteadiness, and frequent falls
  - Visual disturbances, such as blurring or double vision
  - Cognitive difficulties, such as memory problems or changes in mood or behaviour.

#### At diagnosis

- Sensitive communication of the diagnosis to the individual and a supporting family member or friend is essential.
- Offer of appropriate information should be made including:
  - A named professional or first point of contact for information, advice and questions following the diagnosis
  - Local health and social care services and support
  - About PSP and how it might affect the individual and their family
  - The PSP Association and any local support groups.
If the information isn't accepted at diagnosis, the individual should be made aware of how they can access it at a later date.

- The offer of emotional/psychological support should be offered
- Follow up information and appointments should be made
- Comprehensive information should be sent to the GP and the person's key worker
- Referral for a clinical appointment with the person's key worker or member of the multi-disciplinary team should be set up within two weeks of diagnosis.

### During the course of the condition

No two people will experience PSP in the same way, and care and support must be tailored to the individual. However, everyone will require the following core elements:

#### Coordination of Care

- **Access to a key worker or named point of contact** to coordinate the individual’s care and provide support. This might be a specialist nurse, a member of the multi-disciplinary team or other professional with an understanding of PSP, and may change as the condition progresses
- **Liaison and coordination between professionals**, sharing information to avoid duplication and ensure effectiveness of treatments and management of the disease. Multi-disciplinary working is the ideal in effective management of PSP
- **A holistic approach to management** from diagnosis onwards, so that the individual’s impairments are seen in relation to each other and care which is provided by one professional, considers or complements that provided by another
- **Regular reviews as necessary**, following a referral, without discharge, to ensure continuity of care
- **Liaison across health and social care**, and supported care planning, reviewed and referred to by all disciplines
- **Referral to the specialist palliative care team from an early stage** for support and advice on approaching the end of life, and early consideration of advanced decisions, particularly if cognitive and / or communication difficulties present early on in the condition
- **Access to appropriate medication with regular reviews.**
Access to information

- **Early access to information**, in line with the individual's wishes, should be made from diagnosis. If the individual is not ready to receive certain information, their family, friends and/or carer may be more appropriate to share this with.

- A **single/first point of contact** for advice, support and signposting should be offered, ideally through a keyworker.

- A **contact number for initial out of hours support** should be given.

- **Regular signposting** to appropriate information should be made, as impairments and disabilities alter, and as new symptoms present.

- **Early information and discussions around advanced decision-making** should be made once the person has come to terms with their diagnosis, to ensure the individual feels in control of their care and wishes, at a time when they are able to make those wishes understood clearly.

**Support for Carers**

- **Information and support in using a Carer's Plan** from social care should be offered, and ongoing referral made to it by health and social care professionals.

- **Respite from the caring role and domiciliary care** to support the care of the individual at home should be offered, with respite opportunity on a regular short-term basis, or for longer periods.

- **Advice and support** on the condition and how to provide care to the individual is essential.

- **Access to services in the carer's own right**, to manage their own physical and mental health, including bereavement counselling and support following the end of life.

**Access to Equipment**

- **Consideration of appropriate equipment** for specific needs should be considered in line with other needs and/or disabilities.

- **Speed of access to appropriate equipment** to ensure the individual can remain independent for as long as possible is essential. Consider sourcing core equipment prior to immediate need.

Education to professionals

- **Education and information for paid carers** including domiciliary and residential care staff, may be required to address appropriate needs, especially around swallowing, cognition and communication impairments.

- **Clear understanding is required by the social services' representative** for the individual, around the rapidity of progression and complex needs of that person, to ensure access to appropriate, timely support and equipment, access to respite care and other supportive services.

- **Education and support should be given to the GP** from clinicians, neurologists or specialist nurses to ensure appropriate support and understanding, prompt referrals, and effective palliative and specialist palliative care is accessed.
Approaching an advanced stage and at the end of life

- **Careful monitoring in advanced stages of the condition is important**, to ensure coordination of care and access to the right support services. End of life is difficult to identify in PSP, and triggers to consider include:
  - Inability to eat and drink/refusal of PEG
  - Infection which might require hospitalisation but which is refused
  - A fall or a major fracture
  - Rapid and significant weight loss
  - Reduced consciousness level without reversible cause and distinct from cognitive impairment, which may be longstanding

- **Access to specialist palliative care** as appropriate and with early referral to ensure prompt access as required

- **GP palliative care registration** should occur promptly, to ensure continuity of care and access, and in conjunction with the Gold Standards Framework

- **Advanced decisions**, which should have been discussed in earlier stages of the disease, should be consulted on again to ensure the individual's wishes are adhered to, including:
  - Reviewing the individual's preferred place of death
  - Reviewing the individual's wishes regarding hospital admission for infections, Peg decision, resuscitation and organ donation
  - Ensuring that all aspects of care, i.e. physical, psychological, spiritual and social, are considered in line with the individual's wishes

- **Access is available to appropriate support and staff who have been trained in end of life care**, whether in the person's home, in a residential home, hospital or other care setting

- **Access to emotional and practical support should be available for carers and family**, including bereavement counselling and support, if required.
Early Stage (including diagnosis)

IN BRIEF...
Able to walk but falls occasionally; difficulty reading due to gaze; mild vocal changes such as quietening; some changes in mood and reduced levels of social interaction. (see reverse for symptoms)

Key Considerations
PSP is often misdiagnosed. Prompt referral for assessment by a movement disorder specialist at first recognition of possible symptoms is essential. Initial symptoms which may indicate PSP include:

- Problems with unsteadiness, balance and frequent falls (often backwards)
- Visual disturbances, such as difficulty with gaze, blurring or double vision
- Cognitive difficulties, such as changes in mood or behaviour, including apathy and anxiety.

PSP may often present through a fracture clinic, eye specialist, falls clinic or speech and language therapist, and greater awareness of the condition and diagnostic markers within these areas could prompt a more timely diagnosis.

Access to a keyworker is likely to ensure coordination of care and prompt access to appropriate services as they are needed. This role may be fulfilled by a specialist nurse (Parkinson’s or neurology), member of the MDT, consultant, community matron, or GP, and may change as the condition progresses.

The individual may be at an early stage, but different symptoms present and progress at different times and rates.

Aims
To ensure that people with PSP and their families are:

- Given a prompt and accurate diagnosis (including ‘possible’ and ‘probable’)
- Well supported at, during and after diagnosis, including in coming to terms with the condition
- Clearly directed to information and support
- Given details of an identified key worker to support ongoing information and access to services
- Assisted to develop awareness, and supported to adapt to the changes of PSP
- Helped to manage and reduce any symptoms
- Assisted in maximising independence and participation in everyday life

Assessment

- Establishment of accurate diagnosis by a movement disorders specialist, ideally with a special interest in PSP (could include consultant neurologist or geriatrician)
- Assess understanding of person with PSP and their family and provide relevant education
- Evaluate symptoms, impairments and patient’s concerns
- Carry out Care Needs Assessment
- Assess social and financial circumstances and support available (including work and driving).

Management

- Offer information and support delivered at individual’s pace:
  – Signpost to the PSP Association
  – Provide contact details for the individual to initiate contact if they wish
  – Offer contact directly with the individual two to four weeks following diagnosis to provide post-diagnostic support (may be via telephone)
- Outline and discuss support available, including drug treatments
- Discuss development of a care plan
- Identify, refer to, and ensure ongoing support from a coordinator or carer (e.g. a keyworker)
- Ensure early access to:
  – Core multi-disciplinary team (MDT)*
  – Counselling and / or psychological support
  – Local voluntary and support organisations
- Ensure regular access to therapy
- Ensure regular review according to the individual’s need
- Provide information about opportunity to be involved in research

*Core MDT would ideally include physiotherapist, occupational therapist, speech & language therapist, dietitian, specialist nurse, social worker and GP.

Outcomes

- Reduction in distress and acceptance of diagnosis
- Support for person with PSP and their family carer(s) ensuring maximised understanding
- Establishment of care networks - medical, nursing, therapy, social, voluntary
- Coordination and continuity of care and timely referrals due to rapid progression of condition
- Prevention of complications including reduced hospital admissions
- Maximised quality of life

This stage typically might span years 0-1. This document is intended to be used by professionals supporting people with PSP. It is only a guide, and every individual should be treated as such, considered holistically, and supported as they prefer. This guide will not describe every individual’s journey, and everyone will progress at different rates: some slower, some faster than this describes. Above all, an individual, personally tailored approach to care, coordinated and timely, is the ultimate goal.
Typical Symptoms

Ensure equal consideration is given to both motor and non-motor symptoms. A range of symptoms may present to various degrees of severity at any stage, although typical at this stage are:

- Unsteadiness, balance problems and falls
- General slowing down
- Quieting of voice
- Emotional / behavioural changes such as apathy
- Depression
- Anxiety
- Problems with vision
- Fatigue.

Information & Resources for people affected by PSP

Consider providing information on the following:

- PSP Association Helpline, Information and Advisory Service - the first point of contact for people affected by PSP, their families and carers
- PSP Association
- Guide to benefits
- Employment support
- Local exercise opportunities
- Family support
- Regional Driving Assessment Centre, DVLA, Blue Badge Scheme and local transport
- Local equipment suppliers
- Disabled Living Foundation
- Access to Work / Job Centre Plus
- Local voluntary organisations including support groups, and organisations such as Age UK and Carers UK
- Information on counselling, including family and individual counselling
- Brain Bank and opportunities for involvement in research.

Information and Resources for Professionals

- PSP Association, including PSP Specialist Care Advisers for advice and support, PSP 'Standards of Care' and 'Symptom Snapshots' for more in-depth information
- Early recognition that equipment is likely to be needed, and prompt procurement of this will maximise its use for the individual before they progress further.
Mid Stage

IN BRIEF...
Individual walks with aids and limited eye movement makes eating and walking more difficult. There is high risk of falls. The individual will not often speak unless directly spoken to, and speech is only understood by those listening carefully. Behaviour is more impulsive with marked apathy. There is a risk of choking when eating and a high level of supervision is required. (see reverse for symptoms)

Key Considerations
Many people may have already reached this stage before they are diagnosed, and so practice set out in ‘Early’ stage should be considered in addition to ‘Mid’, specifically around information, coordination, and psychological support.

Discussion about advance care planning**, preferred priorities of care and advance decisions to refuse treatment should be addressed early on, as deterioration can be very rapid or sudden and changes in communication or cognition could make these discussions difficult.

Early assessment for inclusion on the Palliative Care Register, and access to specialist palliative care with regular reviews for inclusion ought to occur from here onwards due to the rapidly degenerative nature of PSP.

Risky or impulsive actions and behaviour may cause a significant increase in pressure on carers, and an increase in hospital admissions. The individual may require a high level of supervision at all times and in all care settings.

Education to paid carers and paid support is incredibly important as the individual’s needs become more specific and complex. Particularly consider if appropriate:

• Domiciliary care
• Day care
• Residential or respite care
• Hospice at Home.

Aims
• Symptomatic management and prevention of complications
• Maintenance of function, self-care and quality of life despite advancing condition
• Access to therapeutic intervention from multi-disciplinary team
• Agreed individual / family / professional goals
• Coordination and communication between all professionals
• Open communication about the individual’s wishes in more advanced stages
• Carer support.

Re-Assessment
• Assess symptoms, impairments and disabilities including nutritional status
• Risk assessment regards impulsive or risky behaviour
• Ensure appropriate treatment, medication, services and support are available
• Review Care Needs Assessment, and review and update care plan
• Support carer - review and update carer’s plan and consider respite options
• Assessment against prognostic indicators (GSF) to consider access to specialist palliative care.

Management
• Ensure access to core multi-disciplinary team (MDT)*
• Ensure access to social services
• Ensure timely referrals for prompt access to and provision of appropriate equipment and adaptations due to extremely rapid progression of condition
• Referral to specialist palliative care team if appropriate following assessment
• Early opportunity to discuss Advance Care Planning for end of life
• Facilitate symptom control
• Ensure optimum medication is prescribed and regularly reviewed
• Discuss development of carer’s plan.

*Core MDT would ideally include physiotherapist, occupational therapist, speech & language therapist, dietitian, specialist nurse, social worker and GP.

Outcomes
• Effective coordination of all services and professionals across health and social care
• Prompt access to equipment enabling optimum management and independence
• Optimum symptom control
• Clear goals of therapy and care that are both set and adhered to across disciplines, maximising independence, control and quality of life.

This stage typically might span years 2-3. This document is intended to be used by professionals supporting people with PSP. It is only a guide, and every individual should be treated as such, considered holistically, and supported as they prefer. This guide will not describe every individual’s journey, and everyone will progress at different rates, some slower: some faster than this describes. Above all, an individual, personally tailored approach to care, coordinated and timely, is the ultimate goal.

**Care plans and carer’s plans are only useful if referred to by multiple disciplines and across both health and social care. They should be referred to and updated regularly.
Typical Symptoms

Ensure equal consideration is given to both motor and non-motor symptoms. Any variety and severity of symptoms may present at any stage, although typical at this stage are:

- Frequent falls and increased muscular rigidity
- Slowness of thought and increasing difficulty with recall
- Emotional / behavioural changes such as apathy, depression and / or anxiety
- Problems with vision including blepharospasm
- Swallowing problems
- Communication difficulties, e.g. reduced speech and echolalia
- Constipation
- Sleep disorders
- Fatigue
- Pain.

Information & Resources for people affected by PSP

Consider providing information on the following:

- PSP Association Helpline, Information and Advisory Service - the first point of contact for people affected by PSP, their families, and carers
- PSP Association
- Guide to benefits
- Employment support for carer
- Family support
- Local equipment suppliers
- Disabled Living Foundation
- Blue Badge Scheme and local transport (likely carer driving)
- Respite, breaks, and other means of alleviating demands on carer
- Information on advanced care directives and discussion about end of life
- Information on counselling, for both family and individual.

Information and Resources for Professionals

- Continuing Healthcare Decision Support Tool
- Gold Standard Framework Prognostic Indicators
- Information on advanced decisions and advance care planning
  – Preferred place of care and preferred place of death
  – Preferences for care, e.g. PEG feeding, catheterisation
  – Advanced decisions to refuse treatment
  – Lasting Power of Attorney.
IN BRIEF...
Highly reduced mobility and severe muscle stiffness, requiring a wheelchair or confined to bed; severe communication difficulties including lack of expression but fully comprehensive; high risk of aspiration and pneumonia; likely pain, and periods of sleepiness; functional incontinence and severe social withdrawal. (see reverse for symptoms)

Key Considerations
The individual should already have been placed onto the GP Palliative Care Register and should be considered for access to specialist palliative care. This might include:
• Clinical nurse specialists (Macmillan Nurses)
• Hospice day care
• Hospice residential / in-patient care.
The individual may be in pain, and communication difficulties may prevent this from being understood or managed well. This should be taken into account regarding optimum care.

Education to paid carers and paid support is incredibly important as the individual’s needs become more specific and complex. Particularly consider if appropriate:
• Domiciliary care
• Day care
• Residential or respite care
• Hospice at Home.

Considering the individual’s needs: communication difficulties may mean that preparing questions prior to appointments with professionals should be considered, and difficulty travelling to appointments at this stage may make home visits necessary. Increased support for the individual and their carer may be required regarding emotional responses and fears approaching the end of life.

Aims
• Relieve symptoms and distress in person with PSP and family
• Prevent (and where necessary alleviate problems arising from) complications
• Access to respite and carer support
• Maintenance of dignity and remaining function despite advancing condition
• Supported social interaction and communication as far as possible.

Re-Assessment
• Assess symptoms, impairments and disabilities including pain and nutritional status
• Assess if treatment, medication, services and support are appropriate
  – Critical review of risk to benefit ratio, of all medication
• Assess method of communication with individual as appropriate, increasing liaison with carer
• Review and update care plan and consider transfer to Continuing Healthcare
• Review and update carer’s plan
• Review respite care options.

Management
• Manage symptoms, impairments, disabilities and pain
• Ensure access to core multi-disciplinary team (MDT)* and social services and if necessary transfer across to Continuing Healthcare
  – Offer telephone or home visits if preferred by the individual
• Ensure access to community matron or district nurse for regular monitoring
• Ensure timely referrals for prompt access to and provision of appropriate equipment and adaptations due to extremely rapid progression of condition
• Ensure access to regular assistance and support for carer through domiciliary care
• Ensure appropriate out of hours support to minimise unnecessary hospital admissions
• Ensure access to specialist palliative care as appropriate
• Facilitate symptom control
• Optimise medication and mode of administration according to the individual’s needs
• Support of carer e.g. through respite, domiciliary care, day care
• Management of feeding issues, whether with or without use of PEG
  *Core MDT would ideally include physiotherapist, occupational therapist, speech & language therapist, dietitian, specialist nurse, social worker and GP.

Outcomes
• Maintenance of dignity and support in line with individual needs and preferences
• Maintenance of autonomy as far as possible
• Optimum symptom control, managing risk and disability (e.g. aspiration whilst feeding)
• Support to the patient, carer and family
• Maximised quality of life.

This stage typically might span years 3-6. This document is intended to be used by professionals supporting people with PSP. It is only a guide, and every individual should be treated as such, considered holistically, and supported as they prefer. This guide will not describe every individual’s journey, and everyone will progress at different rates: some slower, some faster than this describes. Above all, an individual, personally tailored approach to care, coordinated and timely, is the ultimate goal.

Continued over
Typical Symptoms
Ensure equal consideration is given to both motor and non-motor symptoms. Any variety and severity of symptoms may present at any stage, although typical at this stage are:
• Immobility
• Severe muscle stiffness (particularly neck and back)
• Weight loss
• Dementia, severe slowness of thought and response, and difficulty with recall
• Functional incontinence
• Emotional / behavioural changes, e.g. apathy, depression and / or anxiety
• Severe problems with vision and eye movement
• Swallowing problems
• Severe communication difficulties
• Constipation and incontinence
• Sleep disorders
• Pain.

Information & Resources for people affected by PSP
Consider providing information on the following:
• PSP Association Helpline, Information and Advisory Service - the first point of contact for people affected by PSP, their families, and carers
• PSP Association
• Guide to benefits
• Family support
• Blue Badge Scheme and local transport (carer driving)
• Local equipment suppliers
• Disabled Living Foundation
• Respite, breaks, and other means of alleviating demands on the carer
• Information on counselling, including family counselling, individual counselling.

Information and Resources for Professionals
• Continuing Healthcare Decision Support Tool
• Risk assessment tools for pressure ulcers
• Universal or locally adapted malnutrition screening tool
• Pain assessment tool i.e. PACSLAC Checklist.
End of Life Stage

**IN BRIEF...**
Severe impairments and disabilities and a rapid and marked deterioration in condition; this stage is usually triggered by a decision not to treat, in accordance with the individual’s previous expressed wishes. (see reverse for symptoms)

**Key Considerations**
Before this stage has been reached, preparation should have been made for the individual’s wishes around death, and any Advance Directives, including decisions to refuse treatment, should have been made.

The individual should already have been placed onto the GP Palliative Care Register and should be considered for access to specialist palliative care. This might include:
- Clinical nurse specialists (Macmillan Nurses)
- Hospice at Home
- Hospice residential / in-patient care.

The previous Advanced Stage lasts for an uncertain period of time but there will come a point where the carer and family, and possibly professionals also, will notice a difference in the individual.

End of life is very difficult to detect in PSP as many of the ‘triggers’ highlighted for other conditions are already being experienced. However, triggers to consider for PSP might be:
- Reduced consciousness
- Inability to eat and drink in absence / refusal of PEG
- Infection which might require hospitalisation but which is refused
- A fall or a major fracture
- Rapid and significant weight loss.

**Aims**
- Relieve distress in person with PSP and family
- Prevent (and where necessary alleviate) complications
- Ensure carer support
- Enable maintenance of dignity
- Comply with patient wishes at the end of life as far as possible
- Comply with Advance Directives including decisions to refuse treatment.

**Re-Assessment**
- Reassessment of capacity to make decisions (if making amends to advanced decisions)
- Review Advanced Directives and Advance Care Plans including decisions to refuse treatments preferences for care and place of death, and organ donation
- Assess support networks for carer.

**Management**
- Ensure access, and clear communication of the individual’s wishes, to all relevant professionals
- Ensure access to community matron or district nurse for regular monitoring
- Ensure regular contact with GP
- Ensure appropriate out of hours support to enable care in preferred setting
- Ensure access to specialist palliative care team and specialist palliative nurse
- Facilitate symptom control and optimum medication
- Maximise comfort and pain relief
- Support of carer e.g. through respite, domiciliary care, Hospice at Home
- Referral of carer to bereavement support and services.

**Outcomes**
- Support during time of distress for individual and family
- Maintenance of dignity and adherence to patient wishes, including consideration of preferred place of care
- Maintenance of autonomy as far as possible
- Optimum symptom control
- Support to the patient, carer and family
- Individual dies in preferred place.

This stage typically refers to the last 6-8 weeks of a person's life. This document is intended to be used by professionals supporting people with PSP. It is only a guide, and every individual should be treated as such, considered holistically, and supported as they prefer. This guide will not describe every individual’s journey, and everyone will progress at different rates: some slower, some faster than this describes. Above all, an individual, personally tailored approach to care, coordinated and timely, is the ultimate goal.
End of Life Stage (Symptoms/Information & Resources)

Typical Symptoms
Any variety and severity of symptoms may present at any stage, and not everyone will experience all symptoms. At this point the individual may also be experiencing co-morbidities, and are likely to be experiencing significant pain.

Information & Resources for people affected by PSP
Consider providing information on the following:
• PSP Association Helpline, Information and Advisory Service - the first point of contact for people affected by PSP, their families, and carers
• PSP Association
• Family support
• Spiritual support and services
• The Queen Square Brain Bank for Neurological Disorders (QSBB) and other donor options
• Bereavement support and services such as Cruse
• Information on bereavement counselling for the carer and wider family.

Information and Resources for Professionals
• Legal guidance regarding advanced directives e.g. ‘The Code of Practice,’ British Medical Association 1995
• Capacity, care planning and advance care planning in life limiting illness - A Guide for Health and Social Care Staff 17 May 2011
• National Council for Palliative Care
• www.endoflifecareforadults.nhs.uk
• Advance Care Plans are not legally binding; Advanced Directives are legally binding. www.goldstandardframework.org.uk/advancecareplanning
# Problems with fatigue and sleeping

<table>
<thead>
<tr>
<th>Impairment - considerations</th>
<th>Minimal Impairment</th>
<th>Moderate Impairment</th>
<th>Severe Impairment</th>
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<tbody>
<tr>
<td>The individual may experience some low-level problems. Considerations include:</td>
<td>Occasional or low-level fatigue which requires minimal intervention</td>
<td>Occasional and low level fatigue Some difficulty in both falling asleep and remaining asleep, and averages 5 hours per night or more.</td>
<td>Consistent fatigue which causes distress to the individual Some difficulty in falling asleep and/or remaining asleep, and averages less than 5 hours of sleep per night.</td>
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| Likelihood of symptom at each stage | At early stages, the individual may still be at work, or living relatively independently, but general activities of daily living are likely to require more effort, causing possible fatigue. | As the condition progresses, sleep patterns often become disturbed, there can be difficulty falling or remaining asleep, or alterations in body clock (awake at night, asleep during the day). Pain, bladder and bowel problems, difficulties with movement (both reduced movement and spasms such as ‘restless leg’) and communication, anxiety and medication side-effects can all impact an individual’s sleep. |

| Interventions and Guidance to consider | Advice on fatigue management and possible support and advice in altering or staggering activities through MDT Support from carer or paid carers to manage some day to day tasks Liaison with social care regards domiciliary support may be appropriate Assessment of sleep using sleep history leading to support and advice A sleep history should be taken by the neurologist/geriatrician if the individual reports difficulty sleeping Support from MDT to advise on sleep hygiene and possible aids to support more restive sleep Possible medication to support sleep Regular visits outdoors to access fresh air can improve sleep cycles. | Advice on fatigue management as appropriate Support from domiciliary care as needed Oral fluid and nutritional intake can be affected. Small but frequent meals should be taken Assessment of sleep using sleep history leading to support and advice A sleep history should be taken by the neurologist/geriatrician if the individual reports difficulty sleeping Support from MDT to advise on sleep hygiene and possible aids to support more restive sleep Referral to a sleep clinic may be necessary, if the individual consents and is able to attend Consider contributing symptoms to sleep disturbance such as breathing or swallowing difficulties Consider support to carer should individual’s sleep patterns cause the carer to become sleep deprived Liaison with social care for support including nightsitters Regular visits outdoors to access fresh air can improve sleep cycles. | Advice on fatigue management as appropriate Support from domiciliary care as needed Oral fluid and nutritional intake can be affected. Small but frequent meals should be taken Assessment of sleep using sleep history leading to support and advice A sleep history should be taken by the neurologist / geriatrician if the individual reports difficulty sleeping Support from specialist nurse or referral to occupational therapist to advise on sleep hygiene and possibly provide aids to support more restive sleep Consider support to carer should individual’s sleep patterns cause the carer to become sleep deprived Liaison with social care for support, including nightsitters Regular visits outdoors to access fresh air can improve sleep cycles. |

Continued over
Problems with fatigue and sleeping

- Where sleep disturbances are experienced, medication type and time of administration should be reviewed to ensure it is not a contributing factor.

- Pain, bladder and bowel problems, difficulties with movement and communication, anxiety and medication side-effects can all impact on individual’s sleep:
  - Pain such as in the neck and back, referred pain such as headache, musculoskeletal pain, or pain caused by co-morbidities may all affect sleep, and understanding and treatment of pain experienced may reduce difficulties with sleep.
  - Difficulties with movement including rigidity can mean that waking in the night to use the bathroom or to change position in bed require carer support and take longer, delaying and increasing the difficulty of falling asleep again.
  - Other movement difficulties such as spasms or ‘restless leg’ syndrome can impact on an individual’s ability to fall asleep and to remain asleep. Medication may be able to minimise this in some people.

- Problems with communication can mean that conveying the reason for awaking or for not being able to sleep can protract the length of time before being able to fall asleep. For example, if the individual wakes with a pain, explaining to their carer that they are in pain and where it is may take some time where communication is impaired.

- Appropriate products and equipment to support sleep are important, for example products to support bladder and bowel problems to ensure this does not contribute to sleeplessness or sleep disturbance, and supportive mattresses for comfort in reclining positions might be considered.

- Support and education to the individual, their carer and any nightsitters regarding ideal postures in bed can support better sleep.

- Anxiety regarding end of life can impact sleep. Clear discussions around end of life preferences and opportunity for counselling, emotional and/or spiritual support may help alleviate some concerns.

References
Golbe PSP Rating Scale (number 7) used to create sleep assumptions. Golbe, L and Ohman-Strickland, P. ‘A clinical rating scale for progressive supranuclear palsy’, Brain (2007), 130, 1552-1565.
## Problems with vision
- including diplopia, photophobia, apraxia, vertical gaze palsy

<table>
<thead>
<tr>
<th>Impairment - considerations</th>
<th>Minimal Impairment</th>
<th>Moderate Impairment</th>
<th>Severe Impairment</th>
</tr>
</thead>
<tbody>
<tr>
<td>The person may have slight problems with vision. Considerations include:</td>
<td>Slight decrease in blink rate possibly causing dry eyes</td>
<td>Difficulty in opening and closing eyes and/or involuntary eye closure possibly causing dry eyes</td>
<td>Difficulty in opening and closing eyes</td>
</tr>
<tr>
<td>• Slight double or blurred vision.</td>
<td>Slower or hypometric eye movement (hypometric refers to inaccuracy in looking at chosen target)</td>
<td>Excessive watering of eyes (notably slower or hypometric eye)</td>
<td>Significantly slower or hypometric eye movement, less than half the speed or accuracy of average expectation</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Difficulty with gaze both upwards and downwards</td>
<td>Difficulty with gaze</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Double or blurred vision and/or photophobia (aversion to bright light).</td>
<td>Double or blurred vision and/or photophobia (aversion to bright light).</td>
</tr>
</tbody>
</table>

| Likelihood of symptom at each stage | Double vision or vertical gaze palsy occasionally present early on and often people experience visual disturbance of some kind early on, even pre-diagnosis. | Vertical gaze palsy is likely from mid-stage onwards, and double vision is reasonably common. Photophobia and eyelid apraxia are found in up to half of cases. Problems with gaze can cause trips and falls. | Vertical gaze palsy is likely from mid-stage onwards, and double vision is reasonably common. Photophobia and eyelid apraxia are found in up to half of cases. |

| Interventions and Guidance to consider | • Early referral to orthoptist/eye specialist | • Regular review by orthoptist/eye specialist | • Regular review by orthoptist/eye specialist |
| • Communication with key worker or MDT regarding possible aids or solutions to make activities of daily living easier | • Occupational therapist to consider and/or educate on interventions. e.g. environmental aids to bring objects into person's visual field | • Eyelid apraxia will require eyedrops. Botulinum toxin may be used to treat blefarospasm and apraxia of eyelid opening. If botox is unsuccessful, surgery to the eyelid may be considered |
| • For dry eyes, artificial tears or eye sprays should be prescribed by GP. Ocular lubricants may be used before the individual sleeps | • For dry eyes, artificial tears or eye sprays should be prescribed by GP. Ocular lubricants may be used before the individual sleeps | • Infections need to be checked for regularly. GP can provide support and prescriptions for this |
| • Medications may affect tearing and should be carefully considered during regular medicine reviews by neurologist, geriatrician, community pharmacist or prescribing specialist nurse. | • Eyelid apraxia may be treated with botulinum toxin | • Liaison with community matron or district nurse may be appropriate |
| | | • Education to family and carer regards possibility of tripping due to gaze, and alteration of environment, i.e. de-cluttering the floor to help prevent falls. |
| | | • Occupational therapist to consider interventions. e.g. environmental aids to bring objects into person's visual field, de-cluttering of floor to help prevent falls. |

*Continued over*
Problems with vision
- including diplopia, photophobia, apraxia, vertical gaze palsy

- Difficulty making eye contact may impact communication. Liaison with a speech & language therapist may be appropriate, as may tailoring communication methods accordingly.

- Watering eyes may be problematic as eyes overcompensate for reduced blink rate. Eye drops are still necessary, however, as cornea is still at risk of ulceration. Watery discharge from eyes only bathes the periphery of the sclera and without the blink mechanism the cornea remains vulnerable to becoming dry.

- If certified vision impaired by the ophthalmologist/orthoptist/eye specialist, information should be provided on talking books and newspapers, and signposting to RNIB.

- Simple alterations to the individual’s environment to avoid trip hazards, movement of certain objects within common view, and other such interventions ought to be suggested if difficulties with vision arise.

- For double vision (diplopia) it is worth considering a temporary eye patch/shield to be worn when watching television or eating meals.

- Prismatic spectacles can be used short term to enable more normal vision, for example watching television with extreme axial rigidity.

- Wrap around sunglasses can ease photophobia.

References
Golbe PSP Rating (numbers 13-17) used to consider severity ratings. Golbe, L and Ohman-Strickland, P. ‘A clinical rating scale for progressive supranuclear palsy’, Brain (2007), 130, 1552-1565
Problems with swallowing and respiration

<table>
<thead>
<tr>
<th>Impairment - considerations</th>
<th>Minimal Impairment</th>
<th>Moderate Impairment</th>
<th>Severe Impairment</th>
</tr>
</thead>
<tbody>
<tr>
<td>The person may have slight difficulty swallowing. Considerations include:</td>
<td>• Difficulties eating, including occasional choking or aspiration, potentially caused by cramming mouth with food • May require small sips of liquids and small mouthfuls of food • Occasional coughs to clear fluid • May experience excess or sticky saliva compounding difficulties.</td>
<td>• Occasional coughing when drinking fluids • Drooling of saliva • Difficulties eating, including occasional choking or aspiration, potentially caused by cramming mouth with food • Excess or sticky saliva may increase swallowing and/or breathing difficulties.</td>
<td>The person finds swallowing increasingly difficult. Considerations include: • Aspiration and respiratory problems such as pneumonia • Choking and asphyxiation • Consider PEG feeding • Excess or sticky saliva may increase swallowing and/or breathing difficulties.</td>
</tr>
</tbody>
</table>

| Likelihood of symptom at each stage | Initial bulbar dysfunction including swallowing problems may present at an early stage for around 14% of people, although these problems are more likely in mid-stage. | Over half of people with PSP will experience difficulty swallowing, often from mid/advanced stage onwards. |

| Interventions and Guidance to consider | Prompt referral to speech & language therapist and coordination with MDT and key worker where appropriate • Possible referral to dietician to ensure nutrition is appropriate and to suggest certain dietary changes • Weight should be recorded initially and at regular intervals to ensure any weight loss is noted • Likely to require supervision from carer during meals, snacks or when drinking, particularly if tending to cram mouth with food • May require additional paid carers during meal times to support carer and guard against choking/aspiration. Liaison with social care needed • May experience minimal breathing difficulties, possibly requiring an inhaler or nebuliser, but unlikely to affect activities of daily living. • Medication may be used to dry up saliva • (use hyoscine with caution as can cause hallucinations). | Regular reviews and management by speech & language therapist and wider MDT, to support safety and efficiency of swallowing and to minimise the risk of aspiration. • Referral to dietician as appropriate to tailor nutrition and food types to guard against weight loss • Fluid intake may need to be monitored if dehydration is suspected • Discussion of future treatment options such as PEG should be held early on, in a sensitive manner • Likely to require supervision/support from carer during meals, snacks or when drinking, particularly if tending to cram • May require additional paid carers during meal times to support carer and guard against choking/aspiration. Liaison with social care needed • Equipment provision through liaison with occupational therapist may be required, such as adapted cutlery and plate guards • May experience difficulty breathing • Regular reviews and management with speech & language therapist and wider MDT, to support safety and efficiency of swallowing and to minimise the risk of aspiration • Liaison between speech & language therapist and dietician to optimise intake and nutrition through diet texture modification, and advice on suitable food choices • Regular visit from community matron/district nurse due to likelihood of pneumonia resulting from aspiration • Likely to require support with feeding from carer, possibly requiring additional support from paid carers at meal times. Liaison with social care needed • Consider PEG feeding if indicated as acceptable to the individual • May experience difficulty breathing. MDT to advise on positioning to ease breathing, relaxation techniques and how to maximise breathing efficiency • Education on optimal breathing positions and possible positional relief through equipment may be useful. | Regular reviews and management with speech & language therapist and wider MDT, to support safety and efficiency of swallowing and to minimise the risk of aspiration. • Liaison between speech & language therapist and dietician to optimise intake and nutrition through diet texture modification, and advice on suitable food choices • Regular visit from community matron/district nurse due to likelihood of pneumonia resulting from aspiration • Likely to require support with feeding from carer, possibly requiring additional support from paid carers at meal times. Liaison with social care needed • Consider PEG feeding if indicated as acceptable to the individual • May experience difficulty breathing. MDT to advise on positioning to ease breathing, relaxation techniques and how to maximise breathing efficiency • Education on optimal breathing positions and possible positional relief through equipment may be useful. |
Symptom Snapshots

Problems with swallowing and respiration

<table>
<thead>
<tr>
<th>Interventions and Guidance to consider</th>
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<th>Severe Impairment</th>
</tr>
</thead>
<tbody>
<tr>
<td>The individual should not be discharged from their speech &amp; language therapist following initial assessment</td>
<td>• Consider supporting individual and carer in ‘assisted cough’ technique through physiotherapist</td>
<td>• If necessary, referral to a respiratory specialist may be useful</td>
<td></td>
</tr>
<tr>
<td>The speech &amp; language therapist should liaise with the MDT* to coordinate care. If there is no MDT, liaison across occupational therapist, dietitian and key worker, as well and social services (particularly if agency or domiciliary carers are involved) should occur from the individual’s initial assessment onwards</td>
<td>• Education on optimal breathing positions and possible positional relief through equipment may be useful</td>
<td>• Provision of equipment and education to carers to manage aspiration at home may be required.</td>
<td></td>
</tr>
<tr>
<td>Education and information sharing with professional carers, as well as family carers, is extremely important. If the individual attends a day centre, has domiciliary care in the home, or is in residential care, it is essential the speech &amp; language therapist provides information and education to those professional carers around swallowing</td>
<td>• If necessary, referral to a respiratory specialist may be useful</td>
<td>• Medication may be used to dry up saliva</td>
<td></td>
</tr>
<tr>
<td>Early, sensitive discussion around later treatments, such as PEG feeding - with provision of appropriate information - should be held with the individual and their family carers</td>
<td>• (use hyoscine with caution as can cause hallucinations).</td>
<td>• If necessary, referral to a respiratory specialist may be useful</td>
<td></td>
</tr>
<tr>
<td>Swallowing should be carefully monitored throughout the course of the condition as recurrent respiratory infections are frequent in individuals with PSP and are commonly associated with respiratory-related deaths</td>
<td>• Frequent infection, aspiration pneumonia and/or breathlessness may all be indicative of a move into the end of life stage. Referral to specialist palliative care should be considered.</td>
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</table>

NB. PEG feeding may be necessary, although should be avoided if the individual is cognitively impaired or likely to be distressed by it. If considered, the patient should be fully consensual, with involvement and coordination across the MDT, and possibly the specialist palliative care team and community matron as applicable. If PEG feeding is an option it should be instigated before weight loss is too severe.

*Core MDT would ideally include physiotherapist, occupational therapist, speech & language therapist, dietitian, specialist nurse, social worker and GP.

References
Golbe PSP Rating (number 32) used to consider severity ratings
Golbe, L and Ohman-Strickland, P. ‘A clinical rating scale for progressive supranuclear palsy’, Brain (2007), 130, 1552-1565
Continuing Healthcare Decision Support Tool #6 ‘Nutrition – Food and Drink’ and #9 ‘Breathing’ Care Domain considered:
• For #6: ‘No needs’ and ‘Low’ for ‘Minimal Impairment’, ‘Moderate’ for ‘Moderate Impairment’ and ‘High’ and ‘Severe’ for ‘Severe Impairment’
• For #9: ‘No needs’ and ‘Low’ for ‘Minimal Impairment’, ‘Moderate and High’ for ‘Moderate Impairment’ and ‘Severe’ and ‘Priority’ for ‘Severe Impairment’
### Problems with cognition and mood
- including neuropsychiatric features: cognitive change (memory, language, disorientation, executive function, flexibility), behavioural disturbance (challenging behaviours, impulsivity, apathy) and mood disorders (depression, anxiety)

<table>
<thead>
<tr>
<th>Impairment - considerations</th>
<th>Minimal Impairment</th>
<th>Moderate Impairment</th>
<th>Severe Impairment</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Impairment - considerations</strong></td>
<td>The person may show some indication of cognitive difficulty, behavioural disturbance and/or mood disorder which, with support and reassurance and/or other interventions, is manageable. Considerations include:  • Occasional difficulty with recall and/or some slowing of thought processes  • Altered behaviour such as impulsivity and/or evidence of apathy (often associated with executive dysfunction)  • Mood changes (including anxiety) having some impact on the individual  • Some withdrawal from attempts to engage in planning, support or activities of daily life.  • Coming to terms with the diagnosis itself can often cause anger, aggression and frustration as well as anxiety and depression.</td>
<td>The person may show some cognitive difficulties, behavioural disturbance and/or mood disorders that require consistent support and/or supervision to ensure their safe environment and quality of life. Considerations include:  • Slowness of thought, difficulty with recall or reduced mental capacity requiring support in making decisions about daily life  • Altered behaviour, impulsive and/or apathetic, affecting the individual and their carers’ quality of life  • Mood disturbance (including anxiety or distress) and/or hallucinations that have an increasing impact on quality of life  • Withdrawal from most attempts to engage in planning, support or activities of daily life.</td>
<td>The person may show cognitive difficulties, behavioural disturbances and/or mood disorders that require constant care and supervision, and possible need for access to immediate and skilled response. Considerations include:  • Slowness of thought, difficulty with recall, mental capacity and/or disorientation, making it impossible for the individual to make appropriate choices and decisions about their daily life  • Altered behaviour, impulsive and/or apathetic, significantly affecting the individual and their carer’s quality of life  • Mood changes including depression and frustration, possibly exhibiting as aggression, which has an impact on the individual and their carer’s life  • Withdrawal from any attempts to engage in planning, support or activities of daily life.</td>
</tr>
</tbody>
</table>

| Likelihood of symptom at each stage | Problems with cognition can occur early on. The recklessness tends to become less of a problem once mobility deteriorates. Apathy and behavioural disturbances usually develop later in a high percentage of people.  Some emotional changes at diagnosis, such as depression or anxiety and difficulty with acceptance, is to be expected. | Cognitive difficulties can arise as a side effect of some medication, as symptomatic of the condition and/or as a reaction to aspects of the condition, and are likely to increase as the condition deteriorates.  Apathy, slowness of thought and recall, mood alteration and withdrawal are likely to increase as the condition progresses.  Slowness of thought may be compounded by increasing difficulties with communication. |

| Interventions and Guidance to consider | • Counselling or psychological support offered early on to support coming to terms with diagnosis  • Testing of memory function and possible referral to memory clinic  • Counselling for family members (individual or group counselling) | • Medication management should be tailored to the individual  • If cognitive impairment could be due to medication, the neurologist/geriatrician/PNS should consider altering medication. A medicines review by community pharmacist may also be considered  • Review by clinical psychologist and/or community psychiatric nurse and referral to psychiatrist | • Medication management should be tailored to the individual  • If cognitive impairment could be due to medication, the neurologist/geriatrician/PNS should consider altering medication. A medicines review by community pharmacist may also be considered  • Review by clinical psychologist and/or community psychiatric nurse and referral to psychiatrist |

**Symptom Snapshots**

- Problems with cognition and mood - including neuropsychiatric features: cognitive change (memory, language, disorientation, executive function, flexibility), behavioural disturbance (challenging behaviours, impulsivity, apathy) and mood disorders (depression, anxiety)
Problems with cognition and mood
- including neuropsychiatric features: cognitive change (memory, language, disorientation, executive function, flexibility), behavioural disturbance (challenging behaviours, impulsivity, apathy) and mood disorders (depression, anxiety)

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<thead>
<tr>
<th>Interventions and Guidance to consider</th>
<th>Minimal Impairment</th>
<th>Moderate Impairment</th>
<th>Severe Impairment</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Discussion about advance care planning, preferred priorities of care and advance decisions to refuse treatment, should be addressed early on. Information should be presented appropriate to the individual’s cognitive ability.</td>
<td>• Referral to community psychiatric nurse or clinical psychologist should be considered to support the family in coping with behavioural or mood changes.</td>
<td>• Involvement of and support to family carer should be considered.</td>
<td>• Involvement of and support to family carer should be considered.</td>
</tr>
<tr>
<td>• Communication difficulties can often be misinterpreted as cognitive decline, and this should be guarded against. In particular, slowness of responses should not, in isolation, be considered to indicate a significant cognitive impairment, and patience should be used in communicating with the individual about their care preferences for ongoing support and Advance Care Plans.</td>
<td>• Early discussion about advance care planning, preferred priorities of care and advance decisions to refuse treatment, should be addressed early on, and information presented appropriate to the individual’s cognitive ability.</td>
<td>• Liaison with occupational therapist regarding daily activities and wider care may be appropriate.</td>
<td>• Regular respite periods for the main carer are important to allow them to continue caring for the person at home.</td>
</tr>
<tr>
<td>• Cognitive disturbances or changes in behaviour should be included in the person’s care plan so that other professionals supporting the individual are made aware and can tailor their treatment and care accordingly.</td>
<td>• Referral to memory clinics may be useful.</td>
<td>• Support for carers to help with understanding and caring for emotional and behavioural changes.</td>
<td>• Liaison with occupational therapist regards daily activities and wider care may be appropriate.</td>
</tr>
<tr>
<td>• Education for carer and domiciliary carers may be useful in managing impulsive or risky behaviour, which could cause the individual harm, such as falls from reckless or impulsive movement.</td>
<td></td>
<td>• Education for carer and domiciliary carers may be useful in managing challenging or unexpected behaviours.</td>
<td></td>
</tr>
<tr>
<td>• It is unlikely that the individual’s behavioural changes will cause harm to those around them, although frustration may manifest as anger or verbal aggression at times.</td>
<td></td>
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<tr>
<td>• Access to psychological support for both the individual and the carer/family should be offered as appropriate.</td>
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<tr>
<td>• If consciousness seems to be reduced, distinct from cognitive difficulties, consider referral to specialist palliative care team, as this may signify a move towards the end of life.</td>
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</tr>
</tbody>
</table>

References
Golbe PSP Rating (numbers 1, 2, and 8-10 inclusive) used to consider severity ratings. (Golbe measures different aspects of cognition: withdrawal, irritability, mentation, disorientation, bradyphrenia, emotional lability). Golbe, L and Ohman-Strickland, P. A clinical rating scale for progressive supranuclear palsy, Brain (2007), 130, 1552-1565
Continuing Healthcare Decision Support Tool Care Domains #1 ‘Behaviour’, #2 ‘Cognition’ and #3 ‘psychological & Emotional Needs’ considered:
- For #1: ‘No needs’ and ‘Low’ for ‘Minimal Impairment’; ‘Moderate’ and ‘High’ for ‘Moderate Impairment’ and ‘Severe’ and ‘Priority’ for ‘Severe Impairment’
- For #2: ‘No needs’ and ‘Low’ for ‘Minimal Impairment’, ‘Moderate’ for ‘Moderate Impairment’ and ‘High’ and ‘Severe’ for ‘Severe Impairment’
- For #3: ‘No needs’ and ‘Low’ for ‘Minimal Impairment’, ‘Moderate’ for ‘Moderate Impairment’ and ‘High’ for ‘Severe Impairment’

Symptom Snapshots
# Problems with bladder and bowel symptoms
- including constipation and bladder control

<table>
<thead>
<tr>
<th>Impairment - considerations</th>
<th>Minimal Impairment</th>
<th>Moderate Impairment</th>
<th>Severe Impairment</th>
</tr>
</thead>
<tbody>
<tr>
<td>The individual may experience some constipation. Considerations include:</td>
<td>Low levels of discomfort affecting the individual intermittently or consistently, but to a low degree.</td>
<td>Constipation that requires treatment via medication.</td>
<td>The individual may experience consistent constipation. Considerations include:</td>
</tr>
<tr>
<td>• None, or very minimal, urinary incontinence (a few drops on occasion).</td>
<td>Some bladder incontinence requiring management and potentially requiring containment on occasion or at night.</td>
<td>The requirement for continual medication.</td>
<td>• Some bladder incontinence, either occasional or more consistent, requiring management by containment during both day and night and possibly requiring a catheter.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Some faecal incontinence.</td>
<td>• Some faecal incontinence.</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>• Possible urinary tract infections.</td>
</tr>
</tbody>
</table>

| Likelihood of symptom at each stage | Constipation is a common symptom of the condition. | Constipation is common and is often a side effect of medication to manage symptoms as well as a symptom of the condition itself. It might be that, as medication increases, symptoms worsen. Functional incontinence may also occur from this point. | Towards the advanced and end of life stages, urinary and faecal incontinence become likely. |

| Interventions and Guidance to consider | • Continence assessment from specialist nurse or district nurse required. | • Review management of constipation through contact with specialist nurse and / or other MDT* professional. | • Ongoing review by a continence nurse is required. |
| | • Assessment from OT to aid functional continence. | • Referral to continence advisor may be appropriate. | • Review management of constipation through contact with specialist nurse and / or other MDT professional. |
| | • May be treated with medication, or lessened by dietary changes. | • Support and education to carer on management as necessary. | • Support and education to carer on management as necessary. |
| | • Support from specialist nurse as part of general appointments. | • Ongoing dietary and fluid advice. | • Monitoring genitourinary health and prevention of urinary tract infections. |
| | • Advice on diet and fluid intake in the prevention and management of constipation. | • Possible referral to dietitian. | • Support and advice from community nurse, including information and education on catheterisation, and support should the individual choose to use a catheter. |
| | • Possible referral to dietitian. | | |

*MDT: Multi-Disciplinary Team
• Catheterisation of individuals who are agitated or cognitively impaired is to be avoided as far as possible

• Timely and appropriate access to products and equipment to support continence management is essential

• Incontinence at night may impact on the individual’s ability to sleep, causing or contributing to problems with sleep and fatigue

• Access to management aids to contain incontinence.

*Core MDT would ideally include physiotherapist, occupational therapist, speech & language therapist, dietitian, specialist nurse, social worker and GP.
# Problems with pain

<table>
<thead>
<tr>
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<th>Severe Impairment</th>
</tr>
</thead>
<tbody>
<tr>
<td>The individual may experience some pain. Considerations include:</td>
<td>Ice-cold sensations in extremities</td>
<td>Ice-cold sensations in extremities</td>
<td>‘Break through pain’ caused by movement from co-morbidities or from respiratory difficulties</td>
</tr>
<tr>
<td>• Ice-cold sensations in extremities</td>
<td>• Pain from injuries caused to skin by repetitive behaviour</td>
<td>• Pain from injuries caused to skin by repetitive behaviour or from pressure ulcers</td>
<td>• Neck and upper back pain which can manifest as headache, pain in shoulders or sensation of a ‘muzzy/foggy head’</td>
</tr>
<tr>
<td>• Pain from injuries caused to skin by repetitive behaviour</td>
<td>• Pain caused by falls, muscle rigidity or difficulties with movement.</td>
<td>• Neck and back pain which can manifest as headache, pain in shoulders or sensation of a ‘muzzy/foggy head’</td>
<td>• Lower back pain, around the vital organs</td>
</tr>
<tr>
<td>• Pain caused by falls, muscle rigidity or difficulties with movement.</td>
<td></td>
<td>• Muscle cramps/spasm.</td>
<td>• Muscle cramps, spasm and musculoskeletal pain</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Likelihood of symptom at each stage</th>
<th>Pain is thought to be more common in people with PSP than previously assumed, but difficulties with communication make understanding the extent and type of pain, and managing it appropriately, very difficult.</th>
<th>At later stages, particularly towards the end of life, it is likely that the individual will experience pain, whether neuropathic, visceral, or somatic due to co-morbidities.</th>
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</thead>
<tbody>
<tr>
<td>Pain may be specifically symptomatic of PSP, or resulting from co-morbidities, falls or treatments.</td>
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</table>

<table>
<thead>
<tr>
<th>Interventions and Guidance to consider</th>
<th>• If possible pain is expressed by the individual or detected by a carer or a professional, prompt assessment should be undertaken by a GP or member of the MDT</th>
<th>• If possible pain is expressed by the individual or detected by a carer or a professional, prompt assessment should be undertaken by a GP or a member of the MDT</th>
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</tr>
</thead>
<tbody>
<tr>
<td>• Possible review and management of pain by a neurologist/geriatrician through medication may be required</td>
<td>• Assessment and management should be holistic with attention to impact of other symptoms, particularly emotional and cognitive</td>
<td>• Management of pain through medication via neurologist</td>
<td>• Education regarding appropriate care and support should be provided to any paid carers responding to an individual experiencing pain</td>
</tr>
<tr>
<td>• Referral to a local pain clinic if required.</td>
<td>• Possible review and management of pain by a neurologist/geriatrician/PNS (analgesia/muscle relaxants/Ldopa/antidepressants/anxiolytics may be required)</td>
<td>• Assessment and management should be holistic with attention to impact of other symptoms, particularly emotional and cognitive</td>
<td>• Discomfort and pain may cause emotional responses which could be mistaken for cognitive difficulties</td>
</tr>
<tr>
<td></td>
<td>• Discomfort and pain may cause emotional responses which could be mistaken for cognitive difficulties</td>
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<td></td>
<td>• Consider support to carer in understanding and managing the individual’s pain on an everyday basis.</td>
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*Continued over*
### Problems with pain

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<tbody>
<tr>
<td></td>
<td></td>
<td>Consider support to carer in understanding and managing the individual's pain on an everyday basis.</td>
<td>Pain management of co-morbidities caused by mobility or bladder and bowel management difficulties should be considered, alongside other pain management (if applicable). The GP and/or district nurse may provide interventions for co-morbidities, with referral to specialist palliative care team if necessary. Mild sedation/analgesia/ anxiolytics may be required at end of life if mentally alert/ anxious.</td>
</tr>
</tbody>
</table>

- Education on appropriate care and support to any paid carers regards responding to an individual experiencing pain
- Communication problems may make understanding 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 person's pain and how to alleviate it
- Medication to alleviate pain should be considered and reviewed as the individual progresses
- Consider the different types of pain and the impact of other symptoms and symptom management on them.
# Problems with movement
- including mobility, rigidity, and balance

<table>
<thead>
<tr>
<th>Impairment - considerations</th>
<th>Minimal Impairment</th>
<th>Moderate Impairment</th>
<th>Severe Impairment</th>
</tr>
</thead>
<tbody>
<tr>
<td>The person may experience minor difficulties with complex and fine motor tasks. Considerations include:</td>
<td>The person may experience more obvious problems with mobility and balance. Considerations include:</td>
<td>The person may be experiencing severe mobility and rigidity. Considerations include:</td>
<td></td>
</tr>
<tr>
<td>• Difficulties with writing, shoelaces and buttons</td>
<td>• Unable to stand and sit safely (may need support and assistance from aid or carer)</td>
<td>• Completely unable to stand</td>
<td></td>
</tr>
<tr>
<td>• Possible tremor</td>
<td>• Impaired postural reactions and balance control and/or impulsive or risky movement leading to increased falls</td>
<td>• Unable to assist or cooperate with transfers and/or positioning</td>
<td></td>
</tr>
<tr>
<td>• Some slowness of movement</td>
<td>• Able to stand but balance unsteady</td>
<td>• Muscle rigidity</td>
<td></td>
</tr>
<tr>
<td>• Able to stand but balance unsteady</td>
<td>• Occasional to frequent falls (often backwards).</td>
<td>• Impaired postural reactions and balance control and/or impulsive or risky movement leading to increased falls</td>
<td></td>
</tr>
<tr>
<td>• Occasional to frequent falls (often backwards).</td>
<td></td>
<td>• Muscle rigidity, limited movement of extremities and/or deterioration in posture, gait and stamina.</td>
<td></td>
</tr>
</tbody>
</table>

| Likelihood of symptom at each stage | Bradykinesia, unsteadiness and falls (often backwards) are common within the first year of (suspected) diagnosis with PSP. Balance reactions are often affected. | Movement problems such as impulsivity, rigidity, reduced movement in limbs and problems with general mobility present at an early stage in approximately two thirds of cases. Tremor affects approximately 1 in 10. With progression, falls increase in regularity. | Movement difficulties advance at different rates according to the individual. Most people will require assistance with transfers and positioning eventually. They may require a specialist wheelchair. Increased support from a carer will be needed as movement is lost. |

<table>
<thead>
<tr>
<th>Interventions and Guidance to consider</th>
<th>• Referral to MDT* for early assessment of home (and workplace), for early falls management</th>
<th>• There should be ongoing and regular review by MDT</th>
<th>• Regular review and ongoing education and support from MDT</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Early and ongoing access to physio and/or appropriate supported exercise classes to maintain and optimise movement and mobility</td>
<td>• Intervention as required is essential to maintain and optimise physical movement</td>
<td>• Education for domiciliary and/or residential carers may be needed from MDT or specialist nurse</td>
<td>• Coordination across health and social care is essential to ensure timely access to appropriate equipment such as specialist wheelchair and hoists</td>
</tr>
<tr>
<td>• Equipment assessment is required to support movement and activities of daily living and walking aids may be needed</td>
<td>• Coordination across health and social care to ensure timely access to appropriate equipment is essential</td>
<td>• Good coordination with community matron and/or district nurse, and specialist palliative care team as appropriate</td>
<td>• Good coordination with community matron and/or district nurse and specialist palliative care team as appropriate.</td>
</tr>
<tr>
<td>• Liaison across health and social care may be needed</td>
<td>• Falls management by MDT and referral to a falls team as necessary</td>
<td>• Increased support to carer to provide the level of supervision required where individuals exhibit impulsive behaviour and therefore risky movement.</td>
<td></td>
</tr>
</tbody>
</table>

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*MDT: Multi-disciplinary Team*
Throughout all stages of the condition and at all levels of severity of symptom:

- The individual should not be discharged from their MDT, who should liaise with social services as appropriate. If there is no MDT, liaison across individual disciplines (occupational therapist, physiotherapist, key worker and social services) following initial assessment is essential.
- Access to services will be more consistent if coordinated by a key worker or case manager.
- Consider telecare such as pendant alarms and pressure mats, particularly if individual lives alone.
- Good coordination across health and social care/MDT is essential particularly between the physiotherapist and occupational therapist.
- Whist impulsive or risky movement is a moderate impairment, individuals exhibiting this require a high level of supervision and constant support from the carer, possibly meaning that a greater level of care support at home is required at this time, and the falls potentially incurred from this behaviour can require a high level of access to a wide range of health and social care services.

NB. If it is felt that equipment may be required in the near future, it might be requested/ordered earlier than necessary to ensure that it is readily accessible at the point it is needed.

*Core MDT would ideally include physiotherapist, occupational therapist, speech & language therapist, dietitian, specialist nurse, social worker and GP.

References
Nath, Ben-Schlomo et al 2003 referred to for likelihood of symptoms
Golbe PSP Rating scale numbers 5 and 18-28 inclusive were used to support severity ratings. Golbe, L and Ohman-Strickland, P. ‘A clinical rating scale for progressive supranuclear palsy’, Brain (2007), 130, 1552-1565
Continuing Healthcare Decision Support Tool #5 Care Domain considered: ‘No needs’ and ‘Low’ for ‘Minimal Impairment’, ‘Moderate’ for ‘Moderate Impairment’ and ‘High’ and ‘Severe’ for ‘Severe Impairment’
### Problems with communication

<table>
<thead>
<tr>
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<tbody>
<tr>
<td></td>
<td>The person can generally be understood although some words are unintelligible. Some considerations are: • Palilalia (can sound like stuttering), reduced speech output • Slurred speech • Hypophonia (quiet voice, particularly when tired) • Problems with vision, making eye contact difficult • Hypomimia (loss of facial expression) • Mobility and restricted eye movement may limit communication aid options • Cognitive problems and apathy may lead to withdrawal or difficulty engaging in social interaction.</td>
<td>The person's speech is often unintelligible and communication is more difficult. Some considerations are: • Palilalia (can sound like stuttering) • Reduced speech output • Slurred speech • Hypophonia (quiet voice, particularly when tired) • Problems with vision, making eye contact difficult • Hypomimia (loss of facial expression) • Mobility and restricted eye movement may limit communication aid options • Cognitive problems and apathy may lead to withdrawal or difficulty engaging in social interaction. • Repetitive speech and verbalisation of 'no' and 'yes' when the opposite is meant.</td>
<td>Communication may be incredibly challenging, and factors contributing to this might include: • Very erratic speech, with few remaining words intelligible • Problems with vision, making eye contact difficult • Difficulty with movement, including rigidity, and lack of movement in extremities • Cognitive problems may lead to frustration, withdrawal or increasing difficulty engaging in social interaction of any kind.</td>
</tr>
</tbody>
</table>

| Likelihood of symptom at each stage | At an early stage, some people can experience initial speech problems (14%), cognitive problems (15%) and visual difficulties, all of which may affect communication and social interaction. | It is likely that 3/4 of people will experience speech problems, often from mid-stage onwards. Differing levels of muscle rigidity, cognition and visual difficulties may impact on usefulness of communication aids. | Speech, muscle rigidity, visual disturbance and cognitive issues are likely to increase as the condition progresses, making communication increasingly difficult. |

| Interventions and Guidance to consider | • Prompt referral to a speech & language therapist and coordination with MDT* and key worker • Consideration of communication aids if appropriate; liaison across health and social care • Liaison with clinical psychologist may be appropriate if cognitive problems are impacting communication • Support and working with carer to encourage alternative methods of communication • Provide information on local groups and opportunities for social inclusion. | • Regular review by speech & language therapist and coordination with MDT and key worker • Consideration of communication aids if appropriate; liaison across health and social care • Liaison with clinical psychologist may be appropriate if cognitive problems are impacting communication • Information on local groups and opportunities for social inclusion • Establish non-verbal methods to support communication that can be used when the individual is tired, or later in course of illness. e.g. thumbs up/down, picture/letter boards. • Prism glasses may aid use of picture/letter boards. | • Regular review by speech & language therapist and coordination with MDT and key worker where appropriate • Consideration of communication aids if appropriate; liaison across health and social care • Liaison with clinical psychologist may be appropriate if cognitive problems are impacting communication • Support to family carers in adapting to predominantly non-verbal communication • Domiciliary care should be given by a small number of familiar carers who know the individual and can communicate with them. |
Problems with communication

• The individual should not be discharged from their speech & language therapist following initial assessment

• The speech & language therapist should liaise with the MDT to coordinate care. If there is no MDT, liaison across occupational therapist, dietitian and key worker, as well as social services (particularly if agency or domiciliary carers are involved), should occur from the individual's initial assessment onwards

• Education and information sharing with professional carers, as well as family carers, is extremely important. If the individual attends a day centre, has domiciliary care in the home, or is in residential care, it is essential the speech & language therapist provides information and education to those professional carers around communication

• Speech aids should be considered holistically and with understanding of the individual's overall impairments. Possible difficulties with gaze and vision, and / or with dexterity in the fingers may make a number of aids unsuitable

• Extra time is required for communication as it deteriorates, and all involved with the individual should consider this and take it into account

• Care should be taken to understand the meaning of what the individual is saying, whether due to difficulty in understanding their speech, or due to a response being verbalised which is not what they intend to say (such as saying 'yes' when 'no' is meant)

• Non-verbal methods should be established to support communication that can be used when the individual is tired or has deteriorated further. e.g. hand movements, picture or letter boards. These should be learnt by core caring staff in order to communicate effectively with the individual as they advance through the condition.

NB. If it is felt that equipment may be required in the near future, it might be requested / ordered earlier than necessary to ensure that it is readily accessible at the point it is needed.

*Core MDT would ideally include physiotherapist, occupational therapist, speech & language therapist, dietitian, specialist nurse, social worker and GP.

References
Golbe PSP Rating (number 12) used to consider severity ratings
Golbe, L and Ohman-Strickland, P. 'A clinical rating scale for progressive supranuclear palsy', Brain (2007), 130, 1552-1565
Continuing Healthcare Decision Support Tool #4 Care Domain considered: ‘No needs’ and ‘Low’ for ‘Minimal Impairment’; ‘Moderate’ for ‘Moderate Impairment’ and ‘High’ for ‘Severe Impairment’.
People with PSP will require access to a number of health and social care services throughout the progression of the disease. To obtain a copy of the Care Pathway for PSP please visit www.pspassociation.org.uk or contact the PSPA Helpline on 0300 0110 122. *Core MDT would ideally include physiotherapist, occupational therapist, speech & language therapist, dietitian, specialist nurse, social worker and GP. ** Movement disorder specialists.
The PSP Association has produced this Pathway of Care Guide to help health and social care professionals provide better services to people living with PSP in a timely and appropriate manner. We would very much appreciate your feedback, which will be used as part of our evaluation process of the Guide in 2014. Please complete the questions below and supply any other comments or information in the space provided.

Feedback Form

1. Are you clear about the aims and objectives of the Pathway of Care Guide?  
   Yes [ ] No [ ]  
   If not, please tell us why in the comment box below.

2. Did the content provide you with information you did not already know?  
   Yes [ ] No [ ]

3. On a scale of 1-10 (1=not very; 10= very) how helpful is the content of:  
   a. The standards of care section?  
   b. The symptom snapshots?  
   c. Best practice in PSP?  
   d. The visual diagram?  
   e. The overall content of the guide?  

4. What benefit will it be in your area of specialism?  

5. Will you recommend the guide to other health and social care professionals you are in touch with as appropriate? If not, why not?  
   Yes [ ] No [ ]

6. Is the guide easy to navigate?  
   Yes [ ] No [ ]

7. How did you hear about the Guide?  

8. Can a member of our team contact you in the near future for more feedback?  
   Yes [ ] No [ ]
Pathway of Care Guide Evaluation Continued

Please provide any other useful information for our evaluation process below.

Your Name

Occupation

Work address

Postcode

Work email

Work telephone

Are you currently supporting someone with PSP/CBD? Yes ☐ No ☐

Have you supported someone with PSP/CBD in the past? Yes ☐ No ☐

We would like to add you to our database and contact you with future news and events about PSP and the PSPA.

If you do not wish to be added to our membership database please tick here: ☐

Would you be willing to help shape the development of our information in the future? Yes ☐ No ☐

For further information please contact:
Carol Amirghiasvand 01327 356137, email helpline@pspassociation.org.uk