**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, which should be considered in management of specific symptoms and holistic treatment of the individual.

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**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 keyworker 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 2-4 weeks following diagnosis to provide post-diagnostic support (may be via telephone);
- Outline and discuss support available including drugs treatment;
- Discuss development of a care plan;
- Identify, refer to, and ensure ongoing support from a coordinator or care (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.

**Outcomes**

- Reduction in distress and acceptance of diagnosis;
- Support for person with PSP and their family carer (s) ensuring maximised understanding.

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**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;
- Quietening of voice;
- Emotional / behavioural changes such as apathy;
- Depression;
- Anxiety;
- Problems with vision;
- Fatigue.

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**Information & Resources for people affected by PSP**

- **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**
- **Information on counselling, including family and individual counselling**
- **Brain Bank and opportunities for involvement in research**

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**Information and Resources for Professionals**

- **Diagnostic criteria:** Litvan, I. et al. Movement Disorders Society Scientific Issues Committee report: ‘SIC Task Force appraisal of clinical diagnostic criteria for Parkinsonian disorders’, Mov Disord 18, 467-86 (2003);
- **PSP Association, including PSP Specialist Nurses for advice and support, PSP ‘Standards of Care’ and ‘Symptom Snapshots’** for more in-depth information.

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*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, which should be considered in management of specific symptoms and holistic treatment of the individual.

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**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.

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This stage typically 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.
Best Practice in PSP - Mid Stage

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

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 is likely to include physiotherapist, occupational therapist, speech & language therapist, dietitian, specialist nurse, social worker and GP.

Outcomes

- Effective coordination of all services and professionals and 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

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

- PSP Association
- Guide to benefits
- Employment support for carer
- Family support
- Local equipment suppliers
- Disabled Living Foundation
- Regional Driving Assessment Centre, DVLA, Blue Badge
- Guide to benefits
- Access to Work / Job Centre
- Disabled Living Foundation
- 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
  - Enduring power of attorney

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.

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.
Best Practice in PSP - Advanced Stage

**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 family support.

**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 & 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 is likely to 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

**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:
- Immobile
- 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**

- 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

**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.

**Outcomes**

- Maintenance of dignity and support in line with individual needs and preferences
- Maintenance of autonomy as far as possible

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.
Best Practice in PSP - End of Life Stage

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 will come to 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 treatment, 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 regards 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

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.

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.

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 comorbidities, and are likely to be experiencing significant pain.

Information & Resources for people affected by PSP

- PSP Association
- Family support
- PALs
- Spiritual support and services
- The Queen Square Brain Bank for Neurological Disorders (Q SBB) and other donor options
- Bereavement support and services such as Cruise
- 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
- National Council for Palliative Care
- Advance Care Plans are not legally binding; Advanced Directives are legally binding. www.goldstandardframework.org.uk 'advancecareplanning'