



# Palliative care in People with **Parkinson's disease**

Guidelines for professional healthcare workers on the assessment and management of palliative care needs in Parkinson's disease and related Parkinsonian syndromes.







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**Disclaimer:** This document is intended as a guideline only. Guidelines will change. Guidelines will not cover all complex clinical cases. Healthcare professionals must use clinical judgement, medical and nursing knowledge in applying the general principles and recommendations contained in this document. Recommendations may not be appropriate in all circumstances and decisions to adopt specific recommendations should be made by the practitioner taking into account the circumstances presented by individual patients and available resources.

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# Introduction

## Strategic context

Palliative care services have progressed considerably in Ireland over the last two decades; a notable advancement came with the publication of the Report of the National Advisory Committee on Palliative Care in 2001<sup>1</sup>.

This report identified the following **principles** of care (p.31):

- “Palliative care is an important part of the work of most health care professionals, and all should have knowledge in this area, and feel confident in the core skills required.
- Primary health care providers in the community have a central role in and responsibility for the provision of palliative care, and accessing specialist palliative care (SPC) services when required.
- SPC should be seen as complementing and not replacing the care provided by other health care professionals in hospital and community settings.
- SPC services should be available to all patients in need wherever they are and whatever their disease.”
- This report also recommends that palliative care should be incorporated into the care plan of patients at an early stage of their disease trajectory, rather than considering palliative care as appropriate only in advanced disease or at end of life.

Although palliative care services have been largely associated with cancer care, it is increasingly recognised, as above, that the principles of palliative care are not just confined to malignancy, but need to be embedded in frameworks of all life-limiting disease<sup>2</sup>. More recently, the Palliative Care Competencies Framework was published which outlines core competences and discipline specific competences for 12 health and social care disciplines, which apply to all life-limiting illnesses<sup>3</sup>.

The role of palliative care specifically in neurological disease in Ireland has been highlighted<sup>4</sup>. A series of Irish research studies have indicated a need for healthcare workers (HCW) to have better training and skills in palliative care for people with Parkinson’s disease<sup>5, 6</sup>.

The aim of these guidelines is to provide practical guidance on the assessment and management of palliative care needs in Parkinson’s disease and Parkinsonian syndromes (PD; see Appendix I for list of abbreviations).

### Development of the guidelines

The Irish Hospice foundation funded the development of these guidelines following a successful grant application. A steering group was established, and a part-time research co-ordinator was appointed in October 2013. Three independent primary research studies were conducted with HCW; people with PD; and families/ carers; to explore palliative care needs in advanced PD. These studies have highlighted a need for HCW to have better training and skills in palliative care for people with PD<sup>5-7</sup>.

In the summer of 2014, an expert advisory group for the guidelines was established, and the first meeting of the group was held in October 2014 (see Appendix II for a list of members of the steering group and advisory committee). The Guideline Developers Manual of the National Clinical Effectiveness Committee<sup>8</sup> informed the development process for these guidelines. The goal was not to create new guidelines, but to adapt existing high quality guidelines to the Irish context. A comprehensive search was performed to identify previously published guidelines for palliative care in PD. No guidelines were found which exactly met this criteria, however a number of documents which are relevant to the aims and objectives of this guideline were located.

The primary documents which were adapted for the current guidelines are:

- NHS England [formerly the National End of life Care Programme] (2010). *End of life care in long term neurological conditions: A framework for implementation*. The National Council for Palliative Care; the Neurological Alliance; NHS National End of life Care Programme.<sup>9</sup>
- The National Council for Palliative Care (2012). *Parkinson's and the last days of life: Consensus statement on the management of symptoms for people with Parkinson's and related conditions in the last few days of life*.<sup>10</sup>

A second comprehensive search was conducted to identify guidelines for symptom management in PD. The guidelines included were published by the following groups:

- National Institute for Health and Clinical Excellence (NICE; 2006)<sup>11</sup>
- Scottish Intercollegiate Guidelines Network (SIGN; 2010)<sup>12</sup>
- Movement Disorder Society (MDS; 2011, 2011)<sup>13, 14</sup>
- American Academy of Neurology (AAN; 2010)<sup>15</sup>
- European Federation of Neurological Societies (EFNS, 2011)<sup>16</sup>

Where recommendations are taken from one of these existing guidelines, the source for the original recommendations (NICE, SIGN, MDS, AAN or EFNS) is referenced. The systems for determining the level of evidence that were used across the guidelines differed slightly but the grade for the recommendation was maintained from the original source (see Appendix III for detailed grade system for each guideline). A systematic review of the peer-reviewed literature was also conducted to identify relevant studies which were published since the publication of these guidelines (this was conducted in March 2015 and reviewed literature published between 2010 and 2015). Studies were included where they were of high quality and were not referenced in previous guidelines. The search strategy is detailed in Appendix IV.

A draft guidelines document was developed following the above rigorous processes, overseen by the advisory group. The draft document was sent for international expert review in July 2015 (reviewed by Professor David

Oliver and Professor Anette Schrag). Minor changes were made following their feedback, and accepted by the advisory group. Finally, the guidelines were distributed for public consultation in Ireland between August and September 2016. A list of the individuals / organisations who provided feedback during the public consultation process is provided in Appendix V. Again, changes were approved by the advisory group. Final editing and proof-reading was undertaken in April 2016. The current guideline was needed as there existed no previous comprehensive overview of palliative care needs in PD. While the existing NHS England<sup>9</sup> document covered end of life care in neurological illnesses, the current guideline expands on this development by adding information specifically related to the care of people with PD. Furthermore the current guideline adds information on the drug and non-drug management of symptoms relevant to the end of life period, based on recent research.

### Background

Parkinson's disease is a progressive neurodegenerative condition, resulting from the death of the dopamine producing neurons in the substantia nigra of the mid brain, and is currently incurable. Thus all treatment is symptomatic, with an average life expectancy post diagnosis of 15 years, although this can vary greatly. For example, being diagnosed with Parkinson's disease at an older age is associated with decreased life expectancy<sup>17</sup>. Parkinsonian syndromes, such as progressive supranuclear palsy (PSP) and multiple system atrophy (MSA) have shorter life expectancies, about eight years after symptom onset or within four years of diagnosis<sup>18</sup>. Parkinson's disease is the second most prevalent neurological disease in Europe, after Alzheimer's disease. It affects 1% of those over 60 years, and 2% over 80 years of age (approximately 12,000 people in Ireland<sup>19</sup>). Because the risk of developing Parkinson's disease increases with age, the fact that more people are now living into old age means that the overall number of people with Parkinson's disease is also rising. The prevalence of Parkinson's disease is expected to more than double by 2030<sup>20</sup>.

### Burden of PD

PD is a substantial burden on the person, and also their families and carers<sup>11, 21</sup>. Considerable research has documented caregiver burden in Parkinson's disease, including depression, stress, strain, fatigue and mortality<sup>22, 23</sup>.

### Economic impact

The economic consequences of Parkinson's disease include both direct and indirect costs. Direct costs include the cost of medicines and medical care and aids/equipment and adaptations. Indirect costs include early retirement or loss of employment for the person with



Parkinson's disease, reduced working hours for carers, and the costs of additional home or institutionalised care<sup>24</sup>.

One study found that the average semi-annual (6 month) cost per patient with Parkinson's disease across four Western European countries was 7442.50<sup>25</sup>. As the population continues to age, the economic burden related to Parkinson's disease will continue to escalate. A more recently published study in the United States<sup>26</sup>, found that the annual utilization of health services and cost for the Parkinson's disease cohort was significantly higher than for a control population. On an annual basis, a person with Parkinson's disease spent approximately two more days in hospital, 43 more days in long-term care institutions, and fill more than 20 more prescriptions than do the controls. The total annual cost is more than double that of the control population, even before adding indirect costs (uncompensated care, productivity loss, etc.).

**Guideline title:** Palliative care in People with Parkinson's disease: Guidelines for professional healthcare workers (HCW) on the assessment and management of palliative care needs in Parkinson's disease and related Parkinsonian syndromes.

**What the guideline covers:** Complex symptom management of Parkinson's disease and related Parkinsonian syndromes, especially in advanced disease.

**What the guideline does not cover:** Symptom management in Parkinsonism (e.g. isolated shuffling gait, medication induced tremor, age related slowness of movement); diagnosis and early assessment; treatment of early Parkinson's disease and related Parkinsonian syndromes.

**Scope:** This guideline is not intended as a policy document. This guideline will form part of any future full Irish guidelines for PD. This guideline will serve as an information document for HCW who may be unsure of the appropriateness of a palliative care approach, or referral to SPC. Readers are also directed to the referral criteria for SPC developed by the National Clinical Programme for Palliative Care, available at <http://www.hse.ie/eng/about/Who/clinical/natclinprog/palliativecareprogramme/Resources/refer%20specialist%20palliative%20care%20services.html>

**Population:** People with idiopathic Parkinson's disease and related Parkinsonian syndromes (PD), their carers, and family members. Parkinsonian syndromes include: Multiple System Atrophy (MSA), Progressive Supranuclear Palsy (PSP), Corticobasal Degeneration (CBD), Lewy Body Dementia (LBD), and Parkinson's Disease Dementia (PDD).

For convenience, Parkinson's disease and related Parkinsonian syndromes will be referred to as 'PD' throughout this document, unless otherwise indicated. Where certain management and treatment may be different for a particular Parkinsonian syndrome, this will be highlighted in the guidelines

**Users:** These guidelines are intended for **all professional healthcare workers** who are involved in the care of people with PD and their carers/families, in any acute, community, or residential setting in Ireland.

#### **Aim and objectives of the guideline**

**Aim:** To provide practical guidance on the assessment and management of palliative care needs in Parkinson's disease.

#### **Objectives:**

- 1 To optimise function and quality of life in people with PD
- 2 To highlight the benefits of a palliative care approach for people with PD
- 3 To provide information to HCW who may be unsure of the appropriateness of adopting a palliative care approach in PD
- 4 To promote access of people with PD to SPC services, when required
- 5 To raise awareness of the prevalence of cognitive impairment / dementia in late PD
- 6 To promote timely care planning and intervention
- 7 To improve the cohesiveness of the approach of the health / social services provided to people with PD, and hence improve the quality of care of the person with PD.

# Palliative Care Approach in PD

The World Health Organisation<sup>27</sup> defines palliative care as:

*“An approach that improves the quality of life of individuals and their families facing the problem associated with life-threatening illness, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial and spiritual.”*

It is important to note that this definition does not refer to any one disease, and the definition can be applied to any “life-threatening illness”. As PD is currently incurable, it is important that a “palliative care approach” is used from the outset – i.e. a focus on assessing and managing symptoms and improving quality of life throughout the course of this life-limiting illness, complementing but not replacing other treatments. This does not necessarily mean that the person with PD needs to be referred to a SPC team or a hospice, just that all HCW are aware that promotion of quality of life is the main goal of care. **Table 1** outlines three levels of palliative care provision.

However, there may come a time in the disease that SPC team input would be valuable. The trigger for this is that the patient has unmet care needs that can't be met by other services. This doesn't depend on disease duration or stage – please refer further to the National Clinical Programme for Palliative Care guidance on palliative care needs assessment: <http://www.hse.ie/eng/about/Who/clinical/natclinprog/palliativecareprogramme/Resources/needs%20assessment%20guidance.html><sup>28</sup>).

It is useful to note that the advanced disease phase in PD may last about 2.2 years<sup>29</sup>, although in PSP, CBD and MSA there can be a more sudden and rapid decline. This advanced phase in Parkinson's disease may be defined<sup>30</sup> by:

- The presence of advanced comorbidity
- Inability to tolerate adequate dopaminergic therapy
- Unsuitability for surgery

This advanced stage should trigger very frequent re-assessments for unmet palliative care needs.

All HCW working with people with PD should be familiar with the Palliative Care Competence Framework, especially as it applies to their specific discipline. The framework describes core competences and discipline specific competences for twelve health and social care disciplines, and is designed to foster inter-professional and inter-organisational collaboration in palliative care provision. The framework can be accessed at: <http://www.hse.ie/eng/about/Who/clinical/natclinprog/palliativecareprogramme/Resources/competence%20framework.html>

## **Dynamic involvement of palliative care throughout the illness**

The progression of PD is highly variable, making planning challenging. It is therefore essential that each person is assessed regularly and their changing needs are managed on an individual basis. There are tools available for assessing palliative care needs in Parkinson's disease, but not all are validated in this



<b>Level 1 - Palliative Care Approach</b>	Palliative care principles should be practiced by all health care professionals. The palliative care approach should be a core skill of every clinician at hospital and community level. Many individuals with progressive and advanced disease will have their care needs met comprehensively and satisfactorily without referral to specialist palliative care units or personnel.
<b>Level 2 - General Palliative Care</b>	At an intermediate level, a proportion of individuals and families will benefit from the expertise of health care professionals who, although not engaged full time in palliative care, have had some additional training and experience in palliative care, perhaps to diploma level. Such intermediate level expertise may be available in hospital or community settings.
<b>Level 3 - Specialist Palliative Care</b>	SPC services are those services whose core activity is limited to the provision of palliative care. These services are involved in the care of individuals with more complex and demanding care needs, and consequently, require a greater degree of training, staff and other resources. SPC services are available within primary care settings, acute general hospital settings and specialist inpatient units.

Table 1. Levels of palliative care specialisation (NACPC: 32)

population. Tools include the Edmonton Symptom Assessment Scale PD (ESAS-PD<sup>31</sup>), Palliative Care Outcome Scale for Symptoms (POS-PP<sup>32</sup>), Functional Assessment of Chronic Illness Therapy Spiritual Wellbeing (FACIT-Sp<sup>33</sup>), Zarit Caregiver Burden Index (ZCBI<sup>34</sup>), and and Needs Assessment Tool-Parkinson's Disease (NAT-PD<sup>35</sup>).

As individual needs vary over time, a model of dynamic involvement of palliative care services should be adopted (see 'model C' in figure 1). A general palliative approach is applicable from diagnosis. General and specialist palliative care might become more involved at times of particular symptoms or psychosocial issues, e.g. pneumonia, marked functional decline, spiritual crisis, as well as at the very end of life<sup>9</sup>. It is important to note that people with PD may have periods where they require SPC input, but can emerge from these to a state at or close to their level of function and quality of life before the episode. This highlights the appropriateness of 'model C' and emphasises that palliative care is not synonymous with end of life.

Another key feature is that SPC input has to be integrated with usual care, with the SPC team working

alongside the usual treating HCW. Critical to this dynamic model of care is good coordination and communication between all professionals involved in the MDT, to maximise quality of life for the person with PD and their family.

### PD and related disorders

The term "Parkinsonian syndromes" refers to a group of diseases that are all linked to an insufficiency of dopamine in the basal ganglia - the part of the brain that controls movement. Symptoms include tremor, bradykinesia (extreme slowness of movement), flexed posture, postural instability, and rigidity, but the extent and severity can vary within the syndromes. Idiopathic Parkinson's disease is responsible for 85% of all Parkinsonian syndromes<sup>36</sup>. Parkinsonian syndromes exhibit the main symptoms of PD, mentioned above. Additional features that are characteristic of the other diseases are as follows<sup>37</sup>:

*Multiple System Atrophy (MSA)*: MSA is characterized by symptoms of autonomic nervous system failure (symptomatic postural orthostatic hypotension, bladder and bowel dysfunction, erectile dysfunction in men, and urinary retention). Other features include increasingly

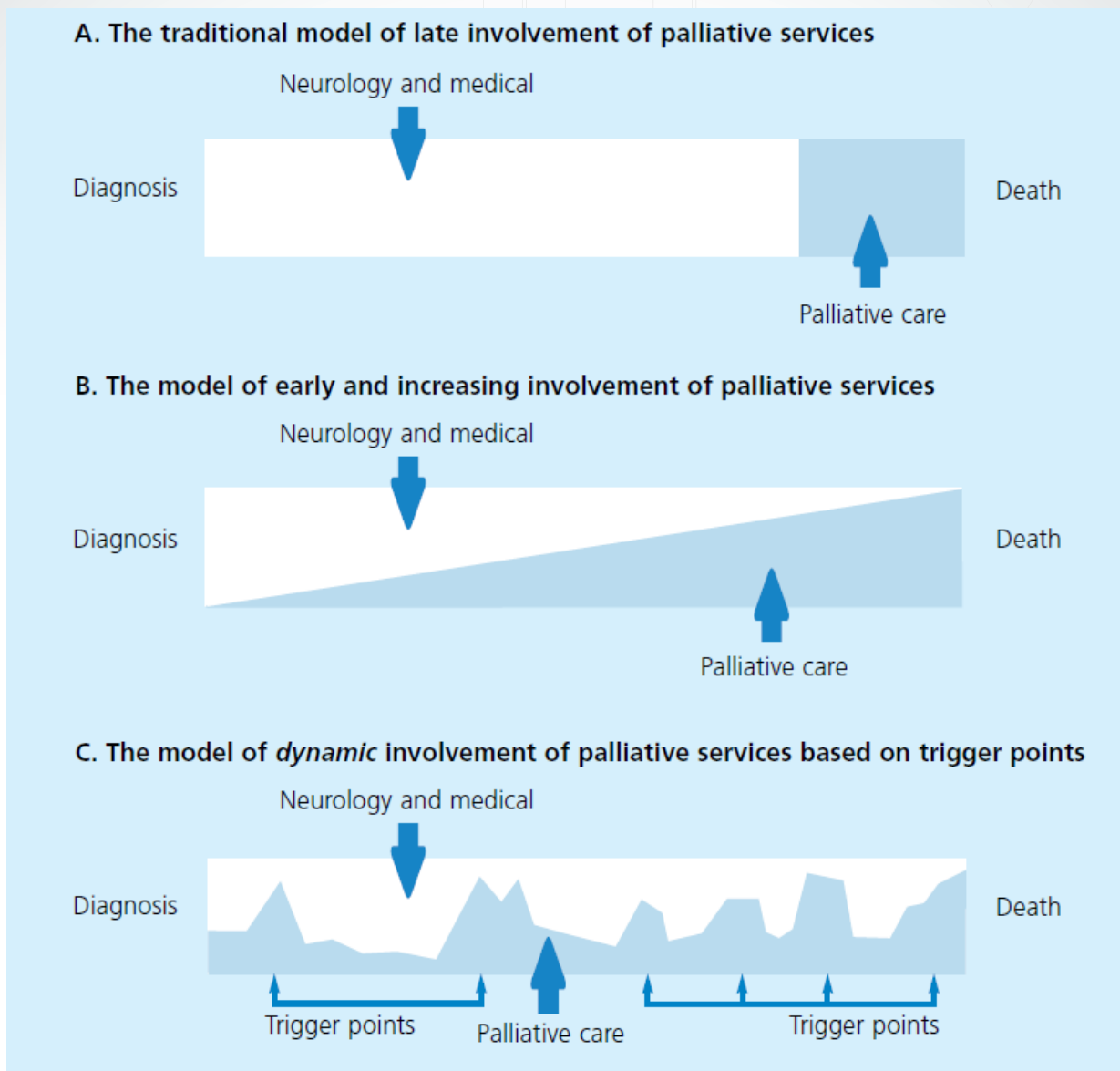


Figure 1. (Reproduced with permission from: NHS England<sup>9</sup>, page 11)

impaired speech and muscle co-ordination, poor balance, and rigidity.

*Progressive Supranuclear Palsy (PSP)*: The cardinal symptoms of PSP include frequent falls, prominent bulbar symptoms especially dysphagia, vertical gaze palsy, and emotional and personality changes.

*Corticobasal Degeneration (CBD)*: The main symptoms of CBD are apraxia (an inability to perform coordinated movements or use familiar objects), pronounced asymmetry often associated with hemiplegia, stiffness that is more severe than classic PD, including marked dystonia, and myoclonus (twitching or jerking) usually in the hand.

*Lewy body dementia (LBD)*: The central feature of LBD is progressive cognitive decline, combined with pronounced fluctuations in alertness and attention, complex visual hallucinations and motor symptoms, such as rigidity and the loss of spontaneous movement. Parkinsonian syndromes including MSA and PSP may be difficult to distinguish initially from PD, but most are usually less responsive to medication and the progression of symptoms is generally more rapid. The prognosis is usually two to four years from diagnosis – people develop considerable disability and require careful assessment and care. This requires a modified palliative care plan, earlier and more intense involvement, and regular review.

# Communication and Care Planning

Effective communication and patient education early in their illness is essential to empower people with PD and their carers to feel they can be an equal partner with HCW in their illness. Equally, effective communication between HCW facilitates consistent and coordinated care. All members of the MDT should therefore develop a therapeutic relationship with clients based on effective communication and information exchange.

Opening dialogue about prognosis and care planning for the future is important and evidence suggests that advanced planning enhances the quality of care<sup>38</sup>. However, some people with PD or their family will not want to engage with palliative care or care planning, at least at first introduction. Equally, HCW need to be sensitive to indicators that a person or family want or need to have such a discussion, and HCW need to be able to facilitate this. However many HCW are reluctant to engage in advance care planning and end of life care discussions with people: fear of causing distress, role uncertainty, appropriate time and lack of confidence<sup>39</sup>. It is important that these challenges are overcome so that the person with PD and their families<sup>40</sup> are:

- given timely information
- can make informed decisions about their future care
- have realistic expectations
- avoid inappropriate burdensome interventions at the end of life

In planning the care of a person with PD, it is obviously important to know the core values of the person. As the majority of people with advanced PD may have communication difficulties and cognitive impairment, these discussions may need to occur earlier in the disease than might occur in other conditions, as the person may not be able to contribute to the discussion in later stages of the disease.

An important point about discussions, conversations and

dialogue with the person with PD, is that these need to be documented and communicated to the relevant MDT members, to avoid multiple questionings about the same issue. However, this is a dialogue over time, and the person's views and wishes may change, so revisiting an issue is sometimes very appropriate.

## Communication throughout the illness

Discussing disease progression and prognosis (particularly in Parkinsonian syndromes with poor prognosis) can be difficult. Individual styles of conversation may vary between people, and clinicians need to be sensitive to this. Thus the following are intended as a guide to conversations, remembering that each person with PD is different (adapted from NHS England<sup>9</sup>):

### In general

- Start difficult discussions at or soon after diagnosis by exploring people's attitudes and views before decisions have to be made and any possible cognitive change has developed
- Recognise that this is a process rather than a single event and will need to be revisited in future consultations
- Ensure that the person / carer understand the reason for referral to palliative care – this might involve a description of the possible role of SPC for their particular symptom at that time. It is important that the person understands that this is not a handing over of care, but a part of their on-going, integrated care



## Planning

- Invite the person to bring someone with them for a planned meeting
- Ensure a private room is available, free from distractions and interruptions, and that the person can remain there after the discussion, to rest and reflect if necessary

## During: Conveying important information

- Convey the information sensitively and at a pace that is manageable for the person; avoid medical jargon
- Allow time for crucial information to sink in; some may need to hear the same information several times

## During: Listening

- Elicit the person's perceptions of their illness
- Keep checking that the person understands what has been said. For example, ask open questions to gauge their level of understanding
- Allow the person to express their feelings, and have these acknowledged

## Follow-up

- Offer literature about their condition to read later, and provide information about sources of support, e.g. PD groups and patient organisations
- Provide a follow-up contact number and arrange a review appointment in the near future for further discussion
- Document the conversation and ensure relevant MDT members are informed

Supporting the individual and their family and ensuring appropriate communication is important in facilitating the individual to come to terms with the reality of having an incurable and progressive neurological disease. Helping individuals and their families understand what is happening now, and how they can best deal with what may happen in the future, is the cornerstone of good neurological care, and is particularly important in PSP, CBD, and MSA where prognosis is poorer. In patients with a Parkinsonian syndrome that involves prominent cognitive impairment, and perhaps poor insight into their own condition, it is important to speak with carers as well as the patient when enquiring about symptoms and when discussing important information.

## Discussing potentially life-prolonging treatments

Many potentially life-prolonging treatments are used in advanced PD without knowledge of the person's wishes. These include treatment of serious infections with antibiotics, and intravenous / subcutaneous hydration, and less commonly enteral nutrition, artificial ventilation, and cardiopulmonary resuscitation<sup>9</sup>.

People's preferences should always be considered. These may differ from the preferences of the family / carer. This needs to be acknowledged and managed skilfully. It is particularly important that a person's family do not feel overly burdened by feeling they are "responsible" for making decisions about medical care.

"It requires great sensitivity to discuss these delicate matters and to discern who does and who does not wish to receive life-prolonging treatment. These difficult discussions are at the heart of compassionate care, respecting the person's autonomy and helping them to make choices in keeping with their own circumstances, attitudes and beliefs. These discussions should start at diagnosis. This is the opportunity to find out about the person's attitudes, before decisions have to be made and cognitive changes have developed. Regular review of care needs is essential."<sup>9</sup>. All HCW involved in the care of a person with PD should be comfortable in discussing care decisions, including life prolonging treatments, within their own competencies. Informal or impromptu discussions can be followed by a formal meeting with more members of the MDT, as appropriate, including the person with PD wherever possible, and relevant carers / family members, so that the person's / carer's questions can be fully answered, family members get to hear the same information, and a plan for future care can be discussed based on up-to-date information on the person's current status.

Suggested prompts for talking about life-prolonging treatments<sup>9</sup>:

*"Has it been helpful in the past to know a bit more about your condition?"*

*"Do you like to know what is happening with your condition?"*

*"Would it be helpful to talk about how this condition may progress from here?"*

*"How have you been coping with things since you found it harder to swallow?"*

*"What you would like to see happen from here?"*

### Discussing advance care planning

Advance care planning is “a term to describe an advance expression of wishes by a person at a time when they have the capacity to express their wishes, about certain treatment that might arise at a future time when they no longer have capacity to express their wishes”<sup>41</sup>. Advance care plans can improve end of life care, improve family satisfaction, reduce stress, anxiety and depression in surviving relatives<sup>38</sup>. Advance care directives will be discussed separately later.

The need for information and the wish to write an advance care plan will vary between individual cases. However clinicians should provide the opportunities to have open and honest conversations – including end of life care discussions – at all stages of the illness<sup>9</sup>.

There may be times when clinicians need to actively encourage advance care planning, e.g. when discussing life prolonging treatments, as above. The difficulties in diagnosing PSP, CBD, and MSA often mean that people can have quite advanced illness and significant communication difficulties at diagnosis and advance care planning may be appropriate soon after diagnosis. It is important to open the dialogue with the person about their disease, including an opportunity for advance care planning if the person wants this, early in these disease trajectories, soon after diagnosis.

Specific issues around care planning in advanced disease / end of life care are discussed later under the heading “Planning Future Care and End of life Care in PD”.

# Multidisciplinary Care

It is important that there is a clear, defined pathway to follow when an individual needs help or advice. Team members may be geographically dispersed so it is good practice to have a single point of contact, i.e. a ‘key worker(s)’. It is essential that the individual, their carers, and the primary HCW involved always know who to contact should a need arise.

It is also important that the MDT is kept updated of relevant changes in the person’s condition or wishes, particularly the GP, who is often the first point of call of the person / carers. A well-functioning MDT will help to ensure that all aspects of the person with PD and their families’ needs are met, in any setting. Excellent communication between professionals is essential to avoid problems such as: individuals receiving conflicting information; individuals and their carers having to repeat information many times; having anxiety and confusion over who to contact if a particular problem arises; and poor communication resulting in inappropriate hospital admissions. See Appendix VI for a list of HCW who may be involved in the MDT.

### Model of best practice

Two integrated palliative care and Parkinson’s disease services, in Scarborough UK and in Alberta Canada, serve as models of best practice in care. Based on their

experiences, it is important for neurology / geriatrics, rehabilitation, and palliative care teams to develop closely co-ordinated working links to support all people with PD from diagnosis to death, including:

- Proper flow of communication and information for people and their families
- A designated point of contact for each stage in the pathway
- A needs assessment identifying the patient’s individual problems

Co-ordinated care in Ireland could be greatly enhanced by electronic care records. In order to achieve the best quality of care, professionals must be able to recognise when their own experience is limited, and when to contact other specialists for support.







# Managing Distress in PD

This section discusses the management of advanced PD, typically the final one to two years of life. Where “end of life” care is referred to, this is intended to mean “care that is provided during the period when death is imminent, and life expectancy is limited to a short number of hours or days”<sup>42</sup>, or sometimes in PD, weeks.

This section will be divided into sections that correspond with the four focus areas of palliative care:

- Physical: e.g. pain, breathlessness, rigidity
- Psychological: e.g. depression, anxiety, cognitive impairment
- Social: e.g. loss of employment, role change
- Spiritual: e.g. religious and non-religious issues, asking ‘why me?’

In reality, these four domains have complex, interlinked relationships (e.g. factors and changes in the social, psychological or spiritual domain may influence the experience of physical symptoms by the person), and they are separated out in this guideline only for structure.

In some subsections of Physical and Psychological Care, symptom assessment and management will be discussed separately for i) advanced disease and ii) end of life care. This occurs where the symptom commonly also occurs at end of life, and / or the assessment / management may be different at end of life. Recognising advanced disease and recognising end of life will be specifically discussed in a later section.

# Physical Care

Management of physical symptoms is an essential element of the holistic care of the individual and their carer / family. Physical symptoms requiring palliative care may include the following:

## Rigidity / Stiffness

The causes of rigidity in advanced PD include disease progression, variability in response to dopaminergic medication and increased intolerance to these medications due to neuropsychiatric symptoms, and in those dying of their PD, decreasing drug efficacy.

The balance between relieving rigidity without causing agitation, hallucinations, or somnolence can be difficult, thus expert Parkinson's advice may be required.

Decisions around patient appropriateness for complex PD therapies such as the continuous infusions therapies subcutaneous apomorphine and jejunal levodopa / carbidopa, and Deep Brain Stimulation (DBS) surgery, will usually have been made prior to this advanced stage of illness.

Potential options include (adapted from NCPC<sup>10</sup> unless otherwise indicated):

- Make sure the person gets their dopaminergic medication on time (if swallow is intact).
- Consider addition of PRN doses of dispersible levodopa / benserazide formulations (i.e. Madopar dispersible)
- If dysphagia is interfering with medication administration, regular levodopa / carbidopa formulation (Sinemet) tablets can be crushed or dissolved in water. Note: crushing or dissolving medications will speed up onset of action but also shorten duration of action, so more frequent / lower doses may be needed
- Consider giving dopaminergic drugs via Percutaneous Endoscopic Gastrostomy (PEG) or via Nasogastric (NG) tube; NG use may be especially appropriate for medication delivery during periods of acute intercurrent illness where there is an expectation of recovery.
- Regular levodopa / carbidopa formulation (Sinemet) tablets can be crushed, but

extended release dopamine agonists, or levodopa, combination medications such as levodopa / entacapone / carbidopa (Stalevo) can't be crushed, and so patients should be converted to the equivalent dose of regular levodopa / carbidopa. A PD expert or pharmacist can give advice about drug equivalencies and suitability of a particular formulation for crushing / dissolving.

- Consider rotigotine transdermal patches if enteral administration of medication is not possible (start low and titrate slowly; seek advice from a PD expert as soon as possible as rotigotine may worsen confusion or agitation in advanced PD). Optimal nursing care and pressure area management is vital in all cases.

**End of life rigidity care:** In the final hours / days of life, if it is impossible to administer dopaminergic medication (oral medications impossible and transdermal / subcutaneous dopamine agonist therapy contra-indicated or inappropriate, e.g. delirium, hallucinations), then midazolam in a subcutaneous infusion may be efficacious, although specific research to support this is currently lacking. Clinical judgement, with expert advice if needed, should be used to determine dose, as midazolam is a potentially potent sedative.

## Pain

It is important not to assume that the pain is related to the PD, therefore optimal assessment and regular review are vital, to determine the precise mechanism of the pain, and hence guide treatment. It is important to identify reversible causes of pain (e.g. retention). Receiving usual dopaminergic medication on time, if swallowing, is also important.

The priority is to formally and regularly assess pain / comfort; the best means of doing this will depend on the person, situation and staff member. In routine clinical

practice, a clinical assessment of pain by a competent HCW may suffice. Visual pain scales may be used for patients with communication difficulties. It is also important to consider non-verbal indices of pain, such as groaning, agitation, tearfulness. Consideration may be given to specific pain assessment tools such as the King's PD pain scale<sup>43</sup> (currently in development), or in patients with PD dementia, dementia pain scales such as the Abbey pain scale<sup>44</sup>, Pain Assessment Checklist for Seniors with Limited Ability to Communicate<sup>45</sup> (PACSLAC), or DOLOPLUS2<sup>46</sup>, bearing in mind that they have been developed and validated in a different population.

If the pain is due to rigidity, see previous section. If not consider both non-pharmacological and pharmacological interventions<sup>9</sup>.

**Non-pharmacological interventions:** In advanced PD, non-drug interventions are also important in controlling pain. These interventions include: nursing care; occupational therapy; physiotherapy; positional / postural changes; pressure-relieving devices; Transcutaneous Electrical Nerve Stimulation (TENS); heat; massage and acupuncture. Physiotherapist prescribed passive range of motion exercises provided by the family may also provide relief by maintaining range and function. However it should be noted that this evidence is limited and extrapolated from other neurological conditions<sup>47</sup>.

**Pharmacological interventions:** The World Health Organisation<sup>48</sup> analgesic ladder was developed for cancer related pain. It is often used as a framework for prescribing analgesic pain relief. This involves initially using simple analgesics, such as paracetamol (Step I), progressing to weak opioids (e.g. codeine, tramadol) for mild to moderate pain (Step II), and then strong opioids (e.g. morphine, oxycodone) for moderate to severe pain (Step III), with other adjuvant analgesics, when required. However, it is increasingly recognised that a low dose of a strong opioid may be preferable to a high dose of a weak opioid in terms of analgesic effect / side effect balance. The EAPC recommendations for opioid use in cancer pain<sup>49</sup> include low doses of strong opioids in Step II, with higher doses of these drugs in Step III. For a person with swallowing difficulties, opioids can be given as oral solutions or subcutaneously / transdermally. For acute pain, parenteral routes will give better analgesic effect than transdermal administration. Difficult to manage pain is an indication to seek SPC input. Most analgesia increases risk of constipation, a common symptom in all PD conditions. Monitoring bowel function and titrating laxatives is important to facilitate optimal absorption of PD medications. Please refer to the NCEC guidelines "Management of Constipation in Adult Patients Receiving Palliative Care" for more information<sup>50</sup>.

Other medication may be needed in PD-related pain, e.g. anti-spasticity drugs. Neuropathic pain is common in PD, and should always be considered. Where a person can describe their pain, important clues to neuropathic pain are sensory symptoms such as numbness or paresthesia, and the quality of the pain (burning or radiating / "travelling"). A detailed neurological examination may elicit abnormal motor, sensory, or autonomic nerve function, suggesting that pain may be neuropathic. Validated tools to differentiate neuropathic pain from other types of pain include subjective tools such as Neuropathic Pain Questionnaire (NPQ)<sup>51</sup>, ID Pain<sup>52</sup>, and PainDETECT<sup>53</sup>; and tools that include examination, such as the Leeds Assessment of Neuropathic Symptoms and Signs scale (LANSS<sup>54</sup>) and *Douleur Neuropathique en 4 Questions* (DN4)<sup>55</sup>, which are more sensitive<sup>56</sup>. None of these have been developed or validated in a PD population. The treatment of neuropathic pain includes anti-convulsant agents (gabapentin and pregabalin) and anti-depressants (amitriptyline, duloxetine).

**End of life pain care:** If pain has not been a problem earlier in the illness it is unlikely to be so as death approaches. A dying person may not be able to report pain directly but if they appear restless or uncomfortable and a reversible cause such as a full bladder or bowel has been ruled out it is appropriate to try an analgesic<sup>9</sup>. Please refer also to the NCEC guideline on Pharmacological Management of Cancer Pain in Adults ([http://health.gov.ie/wp-content/uploads/2016/03/Summary\\_Pharma-Mgmt-Cancer-Pain\\_finalwebv6.pdf](http://health.gov.ie/wp-content/uploads/2016/03/Summary_Pharma-Mgmt-Cancer-Pain_finalwebv6.pdf))

## Dyspnoea

Breathlessness in advanced disease is primarily caused by respiratory muscle weakness. Positional changes and postural supports, and chest physiotherapy and cough augmentation to mobilise secretions, can be helpful. People with PD may also have restrictive lung deficits, due to rigidity / dystonia either in "off periods" or as a result of reduced responsiveness to levodopa. Extreme trunk flexion, forwards or laterally, can compromise lung expansion, exacerbated by vertebral osteoporotic fractures; while muscle wasting in advanced disease can affect diaphragm / intercostal muscle strength. Aspiration can further worsen dyspnoea. Incentive spirometry is said to be helpful for restrictive deficits, but motivation and caregiver fatigue can make this difficult. It is important to look for reversible causes of dyspnoea (super-imposed heart failure, pneumonia). Opioids and / or benzodiazepines are useful for easing the distress of breathlessness<sup>9</sup>. Support to manage the anxiety related to breathlessness is an important part of the treatment of dyspnoea (e.g. patient education on dyspnoea as a symptom, breathing techniques, hand-held fan<sup>57, 58</sup>).



**End of life dyspnoea care:** At the end of life, a person with PD may have a reduced ability to cough, and can accumulate secretions in the upper airways. This can result in noisy breathing which is distressing for families / carers. Firstly it is important to explain to the family why the breathing is noisy, what is being done and what its limitations are. Of note, there are no specific studies in PD, but a Cochrane review of treatments (revised 2012<sup>59</sup>) in adult and children populations found no evidence of benefit of any agent to control established secretions at end of life. However, the guidelines group noted that a lack of evidence of benefit to date does not definitively out rule an actual benefit, and these agents are widely used in clinical practice. Of note, recent NICE guidelines for the care of dying adults in the last days of life<sup>60</sup> recommend a trial of these agents if noisy respiratory secretions are present. The guidelines group suggests early introduction of these agents, with prophylactic intent, at end of life, rather than waiting to begin treatment in established secretions, when effects may be less (GPP). Suitable agents include hyoscine hydrobromide or butylbromide, atropine, and glycopyrrolate, with no clear advantage to any one agent as first line treatment. Patient positioning, suctioning (if appropriate), and avoiding overhydration also form part of the overall management. Dyspnoea can be treated with opioids and / or benzodiazepines. It is important to note that at end of life, these agents do not have any negative impact on respiratory function.

### Communication difficulty

As PD progresses, and particularly in MSA, PSP and CBD, the person's ability to speak and be understood can reduce. This can be a combination of low volume voice and dysarthria. Speech and language therapy can improve communication quality (particularly through voice treatment programmes such as the Lee Silverman Voice Technique [LSVT]), and where necessary explore voice and communication assistive technologies. All people with PD should have an effective method for communication throughout their disease<sup>11</sup>.

### Dysphagia and sialorrhoea

Patients should be referred early to speech and language therapy for assessment and management as dysphagia can occur in up to 80% of people with the condition<sup>61</sup> (GPP). Silent aspiration is also an increasing risk in advanced PD<sup>62</sup>. Further instrumental investigation such as videofluoroscopy may be considered appropriate by the speech and language therapist.

There is insufficient evidence to evaluate swallow rehabilitation in PD<sup>16</sup> though a recent systematic review suggests that Expiratory Muscle Strength Training (EMST) and Video-Assisted Swallowing Therapy (VAST)

may be treatment options for dysphagia in PD<sup>63</sup>. The management of severe dysphagia usually involves regular clinical evaluation of the swallow function and consideration of:

- modifying food and liquid consistency
- modifying medication timing and formulation (see earlier section on rigidity/stiffness)
- introducing swallowing manoeuvres
- co-ordinating meal times with "on" times
- educating and training caregivers (GPP)

Treatment of dysphagia is always multidisciplinary in nature and patient-focussed. The maintenance of good oral hygiene is also important for comfort.

Glycopyrrolate can be useful for the short term treatment of sialorrhoea in PD (MDS<sup>14</sup>, Grade A). There is insufficient data on the efficacy of other anti-muscarinic agents, and they may worsen confusion in advanced PD (this includes hyoscine patches). Botulinum injection of salivary glands, if available, has proven efficacy for sialorrhoea in general populations, and specifically in PD (Grade B), and due to its local effects, can be used in advanced disease without causing cognitive deterioration. Atropine drops can be used sublingually in selected cases where quality of life is affected (GPP). The potential use of anti-muscarinics in a person with existing cognitive impairment should usually be discussed with a PD expert and the person / family should be made aware of the potential for cognitive worsening / hallucinations.

### Artificial nutrition and hydration

Sometimes, a person with PD will develop a potential indication for artificial nutrition and / or hydration. If no preference has been expressed and the individual is not well enough to make a decision, the MDT must decide what is in their best interests, weighing up the advantages and disadvantages. Whilst artificial feeding (via gastrostomy, inserted either as a percutaneous endoscopic gastrostomy [PEG] or radiologically-inserted gastrostomy [RIG]), or hydration, can be beneficial in selected cases, where there is a reversible element to the person's illness, it may not be appropriate, for example where the person has a limited life expectancy, poor quality of life or advanced dementia (please refer to later section on "Recognising advanced disease and limited life expectancy").

Therefore there are a number of key factors to be considered before either treatment is administered (EPDA<sup>64</sup>), including:

- respecting the individual's wishes and acknowledging those of the carer and family
- allowing the individual, if competent, the right to decide whether they want to eat or not
- acting in the individual's best interests
- maintaining wellbeing and the best possible quality of life
- providing supportive care
- minimising the effects of malnutrition
- minimising any side effects of treatment

When artificial feeding is an option, key factors that need to be taken into consideration include (EPDA<sup>65</sup>):

- hunger
- whether nutritional supplements are already being given
- if a desire to live or die has been expressed
- whether it is the individual's wish that nutritional support be given
- whether such support might increase the risk of complications

Artificial hydration might improve some PD symptoms such as delirium, nausea, constipation and orthostatic hypotension but it might not alleviate symptoms such as a dry mouth or thirst, and in some cases it might worsen restlessness and incontinence. Of note, a Cochrane review in 2008<sup>66</sup> found that there were insufficient good quality studies to make any recommendations regarding the use of artificial hydration in palliative care patients (not specifically in PD).

**Artificial nutrition and hydration at end of life:** If a person already has a gastrostomy tube fitted, when dying it is often appropriate to reduce the volume of feed, or even to stop because of the body's diminishing ability to handle fluid or nutrition. Continuing nutrition at this stage may even exacerbate some symptoms such as respiratory secretions. A systematic literature review of artificial nutrition and hydration in the last week of life in cancer patients found little evidence that these improved symptoms or comfort<sup>67</sup>. Withdrawing feeding or fluids requires careful communication with family and carers, who may interpret these changes as a cause rather than a result of dying.

## Pressure ulceration

In the advanced stages of PD many people become immobile. This places individuals at risk of pressure ulcer development, and an assessment of risk for pressure ulcers should be a priority. Most pressure ulcers occur over a bony prominence. Dystonia and contractures of the limbs may cause pressure ulcers in more unlikely places. HCW should assist the carers in understanding how to move such physically dependent people and handle their relative safely. An occupational therapy assessment may assist in improving posture, and seating and sleeping positioning and cushioning (e.g. wheelchair review, pressure relieving cushions, gel heel / elbow / ear protectors). Skin pressure pain from immobility may be relieved by opioids. Pressure relieving devices for both the bed and chair should be used.

Proactive diagnosis particularly in the wheelchair bound patients is essential. At every physician appointment, assessing skin integrity is important, especially examining the buttocks for those in wheelchairs, and lower back for those with malnutrition (GPP).

## Nausea

It is important to first consider any underlying causes (e.g. dehydration, constipation, infection, drug causes). Nausea due to dopaminergic medications may be lessened by taking the medications with food (although absorption may be affected). Many routine drugs used for nausea and vomiting, e.g. metoclopramide, cinnarizine, and prochlorperazine, have antagonistic effects on dopamine receptors. They have a very high risk of worsening Parkinson's symptoms and should not be used. Antiemetics which are least likely to cause side-effects in PD are (NCPC, 2012<sup>10</sup>; EFNS late<sup>16</sup>)

- Domperidone, which can be used for gastric stasis type vomiting, can be given orally (oral suspension available), or via suppository. As it blocks peripheral dopamine conversion, it can help dopaminergic medication-related nausea (EFNS late). NB: domperidone has a HPRa warning of cardiac events (May 2014) and so patients, if appropriate for their cognitive status, should be counselled about the risks and benefits of domperidone before treatment, and this conversation should be documented. Please see the following for more information: <https://www.hpra.ie/docs/default-source/publications-forms/newsletters/drug-safety-newsletter-issue-no-61-may-2014.pdf?sfvrsn=8>

- Ondansetron may have a use for constant intractable nausea, but as a selective serotonin 5HT-3 receptor antagonist, it is used mainly where there is disruption to the mucosa of the gut e.g. after chemotherapy or radiotherapy. It is recommended by the EFNS<sup>16</sup> as a second line agent in PD and can be given by various routes. It can cause troublesome constipation.
- Cyclizine can also be considered. However cyclizine may worsen parkinsonism, and should be used with caution.

## Fatigue and sleep disorders

Fatigue can occur at any stage of PD, and sleep disorders may affect up to 90% of people with PD<sup>68</sup>. Excessive somnolence can be particularly challenging for the individual and their families and carer. It can also represent a challenge to nursing staff concerning medication management, hydration, and nutrition. NICE guidelines<sup>11</sup> recommend that good sleep hygiene should be advised in people with PD with any sleep disturbance and includes:

- avoidance of stimulants (for example, coffee, tea, caffeine) in the evening
- establishment of a regular pattern of sleep
- comfortable bedding and temperature
- provision of assistive devices, such as a bed lever or rails to aid with moving and turning, allowing the person to get more comfortable
- restriction of daytime naps (e.g. suggestion that naps are short and earlier in the day)
- advice about taking regular and appropriate exercise to induce better sleep

Drug interventions can also be considered. First, all medication should be reviewed, specifically to avoid any drugs that may affect sleep or alertness, or may interact with other medication (e.g. selegiline, antihistamines, H2 antagonists, antipsychotics and sedatives). Input from pharmacy may be helpful if the person has multiple comorbidities to try to reduce overall medication burden and interactions.

For example, dopaminergic drugs can cause insomnia, as can selegiline, so night-time doses may need to be taken earlier. Conversely, if poor sleep is due to rigidity and inability to turn, dopaminergic medications, including controlled release preparations, may be helpful. Thus, a careful history of the pattern and causes of poor

sleeping is vital. The person / carer should be questioned for restless leg symptoms, nocturia, obstructive sleep apnoea, Rapid Eye Movement sleep disorder, etc.

Modafinil may be considered for daytime hypersomnolence in people with PD (EFNS late<sup>16</sup>, level B; NICE<sup>11</sup>, GPP; SIGN<sup>12</sup> guidelines did not recommend Grade A). Two more recent small studies showed a trend towards, and a significant effect, respectively, in reducing excessive daytime sleepiness. A meta-analysis in 2013<sup>69</sup> (four studies, excluding the most recent positive study) found an overall benefit (overall Grade A evidence). Of note there has been no benefit shown in the symptom of fatigue. Thus, based on the most up to date evidence, we recommend a short trial of modafinil for hypersomnolence, with discontinuation if no effect. If not familiar with this medication, we recommend expert advice is sought.

Methylphenidate may also be considered in patients with fatigue (AAN<sup>15</sup>, Grade C).

## Orthostatic hypotension

Orthostatic hypotension affects about 30-58% of people with PD<sup>70</sup>, particularly those with MSA, and can severely compromise quality of life. Notable causes include poor intake of fluids, side-effects of general medications such as anti-hypertensives, antidepressants, diuretics, other medical conditions such as cardiac dysfunction, and side-effects of all PD medications especially dopamine agonists.

General measures to combat orthostatic hypotension (adapted from EFNS<sup>16</sup>) should include:

- Avoid aggravating factors such as large meals, alcohol, caffeine at night, exposure to a warm environment, volume depletion, and drugs known to cause orthostatic hypotension, such as diuretics or antihypertensive drugs, tricyclic antidepressants, nitrates, alpha-blockers used to treat urinary disturbances related to prostatic hypertrophy. Levodopa, dopamine agonists, and MAO-B inhibitors may also induce orthostatic hypotension.
- Increase salt intake in symptomatic orthostatic hypotension.
- Head-up tilt of the bed at night (30 - 40°) may be helpful.
- Wear full leg length elastic stockings and abdominal binder if person can tolerate.



- Exercise as tolerated.
- Introduce counter-manoevres to prolong the time for which the patient can be upright (leg crossing, toe raising, thigh contraction, bending at the waist).
- In some patients, hypotension occurs only postprandially so it is helpful to warn the patient about this.

Drug therapy can include midodrine (EFNS<sup>16</sup>, Gradel A); fludrocortisone (EFNS<sup>16</sup>, GPP, risk of side effects). However, there is limited evidence for these drugs. A PD expert should be consulted when considering such drugs.

### Summary of physical care

PD causes many motor and non-motor symptoms throughout the disease, and the assessment and management of these can sometimes be difficult, particularly in advanced disease. If initial assessment and treatment attempts have not improved symptoms, it is important to re-assess the nature and cause of the symptom, and also to seek expert help. Clinicians with expertise in PD and SPC teams, often working together, can work with the primary physician and MDT to improve symptoms and comfort.

# Psychological Care

It is important to highlight that many psychological symptoms in PD may be a manifestation of an “off-state” (i.e. a period of low blood-brain dopamine levels, typically associated with rigidity and tremor as well as non-motor symptoms). For example, people may feel depressed or anxious transiently, improving with their next medications. This is different to pervasive symptoms that persist for days, weeks or months. Off-state symptoms should be aggressively targeted by PD medication review. Equally, visual hallucinations and delusions are often precipitated or worsened by PD medications, and again review of PD medications is a key component of their management.

### Cognitive impairment and dementia

As the PD progresses, individuals will invariably experience increasing difficulty with working memory, problem-solving ability, and information processing speed. Dementia is a common feature of later disease, with prevalence of 24-31%<sup>71</sup>. Treatment options for dementia include the acetylcholinesterase inhibitors, rivastigmine (MDS<sup>14</sup> conclusion efficacious; EFNS<sup>16</sup> Late Grade A; AAN<sup>15</sup> Grade B) and donepezil (EFNS<sup>16</sup> Late Grade A ; AAN<sup>15</sup> Grade B). Rivastigmine is available as a transdermal patch, which may be advantageous for compliance. Memantine can be considered for use

if cholinesterases are not tolerated (e.g. worsening tremor), as there is some accruing evidence for its effectiveness (EFNS<sup>16</sup> Grade C). Clinical (neuro) psychology and / or occupational therapy assessments can provide information on the nature and level of impairment present, and guide cognitive re-training and interventions to facilitate adaptation of daily routines, to maximise independence. Carer education is an important, allied role.

### Visual hallucinations

Visual hallucinations are common in people with PD,

and it is important to note that they may not need to be actively treated if they are well tolerated by the patient and carer (NICE<sup>11</sup>, GPP). Where the person is distressed, or there are other features of psychosis, consideration should be given to withdrawing gradually antiparkinsonian medication that might have triggered the psychosis in people with PD (NICE<sup>11</sup>, GPP).

Medications should be withdrawn in the following order: Anticholinergics, amantadine, MAOB inhibitors, COMT inhibitors, dopamine agonists, and then finally, minimize levodopa (EFNS<sup>16</sup>, GPP).

Typical antipsychotic drugs (such as phenothiazines and butyrophenones) should not generally be used in people with PD because they exacerbate the motor features of the condition (NICE<sup>11</sup>, GPP). Atypical antipsychotics may be considered for treatment of psychotic symptoms in people with PD, particularly quetiapine (AAN<sup>15</sup>, Grade C), although the evidence base for their efficacy and safety is limited. Patients with Lewy Body Dementia (LBD) will often present first with cognitive issues, and visual hallucinations / fluctuations in cognition are a hallmark of this disease. Particular care needs to be taken in using antipsychotics in patients with LBD as 50% show neuroleptic sensitivity reactions.

Clozapine at lower doses may be used in the treatment of psychotic symptoms in PD, but regular blood monitoring is required due to the propensity for causing agranulocytosis (NICE<sup>11</sup>, Grade B; MDS<sup>14</sup>, Grade A). Donepezil (EFNS<sup>16</sup> late, Grade C) and rivastigmine (EFNS<sup>16</sup> late, Grade B) may be beneficial for PD psychosis in the context of dementia.

NB sudden onset or worsening of hallucinations, especially accompanied by fluctuating cognition, attention and consciousness level, or deteriorated function, should raise the possibility of delirium (see later section).

## Depression

Up to 60 percent of people with PD experience mild or moderate depressive symptoms, both as a direct disease component and as a result of reduced function, loss of role, and reduced quality of life. It can be difficult to diagnose mild depression in people with PD as some symptoms of depression can simulate symptoms of Parkinson's - such as feeling tired, lacking in energy and sleep and night-time problems. Clinicians should have a low threshold for diagnosing depression in PD (NICE<sup>11</sup>, Grade D).

Mild depression may be helped by complementary therapies, and adjustments to routine such as getting good sleep, improved diet and better exercise; counselling may also be beneficial (GPP).

Moderate to severe depression in early / moderate severity PD may also be helped in the short-term by cognitive behavioural therapy (Grade B). Antidepressants, mainly selective serotonin reuptake inhibitors (SSRIs), can be used to treat moderate or severe depression. SSRIs are considered to have "acceptable risk" for the treatment of depression in PD (MDS<sup>14</sup>) and are commonly used clinically. A recent trial<sup>72</sup> comparing paroxetine, venlafaxine and placebo in patients with predominantly moderate PD (Hoehn and Yahr stages 2 and 2.5) found both active treatments superior to placebo (Grade B). Potential interactions with dopaminergic medications need to be considered. It is also useful to map depressive symptoms to medication timing and other "off" symptoms, as psychological symptoms including depression and anxiety are common during "off" periods. In this case, adjusting dopaminergic therapy may be helpful. Mirtazapine may worsen nightmares but its sedative effects at night are sometimes useful (GPP).

Pramipexole is a dopamine agonist which can be considered efficacious for the treatment of mild to moderate depression in PD (MDS<sup>14</sup>, Grade A). Of note, the most compelling study only included patients with mild-moderate PD (maximum Hoehn and Yahr stage 3), and patients with motor fluctuations were generally excluded from the studies.

## Anxiety

Up to 40% of people with PD may experience anxiety, including generalised anxiety disorder or panic attacks, and anxiety symptoms are frequently comorbid with depression<sup>73</sup>. People with PD may worry about their symptoms worsening, and may worry about the future. Anxiety in PD can often be a wearing off symptom, and this should be assessed. SSRIs may be effective if anxiety is severe, although there are no randomised controlled trials in people with PD. Complementary therapies such as massage, yoga or gentle exercise may be beneficial (GPP). Counselling or cognitive behavioural therapy can also be effective in dealing with anxiety (GPP).

Drug therapy for anxiety also includes benzodiazepines (lorazepam, alprazolam, midazolam and diazepam). However, benzodiazepines may exacerbate falls and are pro-delirigenic, so the risk / benefit must be carefully weighed up.

## Delirium

Delirium is an acute confusional state characterised by altered consciousness, and impaired attention and general cognitive function, which fluctuates in severity over minutes, hours or days<sup>74</sup>. Risk factors for

delirium include underlying cognitive impairment / dementia, older age, medical illness, hip fracture, and psychoactive medications<sup>75-77</sup>. Thus, people with PD are often particularly at risk of delirium when unwell, and HCW need to be particularly vigilant and not assume hallucinations or psychotic features are due to the PD syndrome.

Delirium diagnosis may be particularly challenging in advanced PD, and requires assessment by a clinician with expertise in delirium diagnosis. However, when delirium is suspected, screening for reversible causes of delirium should be performed without delay, including ruling out urinary tract infections or retention, pneumonia, constipation, metabolic abnormalities, pain, medication toxicity, and, where indicated, intra-cranial events<sup>78</sup>.

There is no specific research into delirium treatment in advanced PD, but anti-psychotics and acetylcholinesterases are the usual treatment for delirium (NICE<sup>11</sup> guidelines) and would be expected to also work in PD. As per the treatment of hallucinations discussed earlier, quetiapine is a mild antipsychotic which does not usually worsen PD motor symptoms (although it must be used with extreme caution in LBD). There is no research into the use of clozapine in delirium treatment. Although benzodiazepines have been historically used for the treatment of delirium and do not worsen motor symptoms, they do not treat the underlying psychotic thinking in delirium, and may in fact precipitate delirium<sup>79</sup>, so their use needs to be carefully considered and discussed with an appropriate expert.

**Agitation / delirium at end of life:** Some degree of delirium has been reported in at least 80% of dying people and can give rise to restlessness or agitation. It is important to try to differentiate delirium from simple anxiety, although this can be particularly difficult at end of life in PD if the person's communication ability and motor function is diminished. If delirium is likely, it is important to consider reversible causes, as appropriate

for the setting and individual's status (e.g. an enema for constipation may no longer be appropriate if death is anticipated within hours). A quiet, non-stimulating environment and staff skilled in caring for a person at end of life are important components of care. If a person can swallow, consider quetiapine for delirium. Other antipsychotics are available for administration by other routes, but may worsen rigidity at a time where a person is often unable to swallow dopaminergic medications, and this must be carefully considered.

Benzodiazepines are frequently used at end of life for agitation/delirium, and they have anxiolytic properties. There are also theoretical benefits in PD due to their muscle relaxant properties, at a time where a person may not be able to take dopaminergic medications for rigidity<sup>9</sup>. There is some concern that they may paradoxically precipitate / worsen delirium<sup>79</sup>, although this has not been studied at end of life care or specifically in a PD population. Some degree of delirium has been reported in at least 88% of dying people and can give rise to restlessness or agitation.<sup>79a</sup>

A recent Cochrane review did not find sufficient evidence to support any palliative pharmacological sedation in terminally ill adults during their last few days of life<sup>80</sup> (not specific to PD), but the guidelines group do note that pharmacological sedation is commonly used in clinical practice and is recommended in many general end of life care guidelines. The PD palliative care guidelines group recommends expert input, ideally from a team with joint PD / palliative care expertise, particularly if initial treatment attempts have not relieved the person's distress.

Terminal agitation / delirium can be very distressing to the person's family. They should be supported by a team with skill in end of life care. Ongoing communication is important to help them understand the rationale for a decision to administer sedating medication, as well as the potential consequences.

# Social Care

As someone faces the diagnosis and then the progression of a life-limiting condition such as PD there will be many psychosocial issues that come to the fore, including (adapted from NHS England<sup>9</sup>):

- Fear of the disorder. It may be unknown to the person and their family, or there may be a family history of the disease, with memories of these experiences. Either may be a frightening prospect.
- Fear of the future, including fears around deterioration, dependency and dying.
- Fears for the family, including concerns about how their partner will cope with the death or being alone in bereavement.
- Fear of palliative care. Amongst the public (and HCW) there are common misperceptions about palliative care, firstly that it is applied only at the end of life; and second, that it is synonymous with late-stage cancer care. A reminder of the focus on the quality of life of the person with PD and an explanation of the model of dynamic involvement of palliative care throughout the illness would be helpful as would referring the person with PD and family carers to the resources to the AIIHPC's Palliative Hub (see Appendix VII for list of relevant websites).
- Losses of independence and ability to undertake day-to-day activities as a parent, spouse, sexual partner or participant in sport or other leisure activity.
- Fears of losing abilities, mobility, personal care, feeding, toileting, sexual function.
- Practical issues of finances, housing, making a will and ensuring care for dependants.
- Cognitive changes: as the person loses cognitive ability they may initially be aware of this and fear the progression and loss of awareness and brain function.
- Multiple losses for both the person and their family involving many of the items listed above. This, and the changing family roles involved, may be profound.

Advice and support may be necessary, from counselling and social work support in coping with fears and losses through to financial planning and social services provision. Referral to occupational therapy and physiotherapy may be beneficial to promote independence, mobility, and continued participation in activities. Meaningful occupation is very important for people who are losing their life roles and may help to maintain their sense of personhood. Referral to clinical (neuro)psychology services can be helpful in cases of difficult or complex adaptation to the challenges linked to disease progression. Integrating systemic, psychological and neuropsychological data may allow the MDT to understand and mitigate barriers to engaging with patients whose care has been challenging to date.

HCW can refer carers to PD advocacy groups including the PAI, Move4Parkinson's, Progressive Supranuclear Palsy Association Limited (PSPA Ireland), the Multiple System Atrophy Trust. Individual support may also be required. The websites for these organisations are included in the list of relevant websites in Appendix VII.



# Spiritual Care

Palliative care takes a holistic view of the patient and family. This includes addressing the spiritual care needs of person with PD. The exact definition of spiritual care is not widely agreed. One definition of spirituality is ‘a personal search for meaning and purpose in life’<sup>81</sup>. It is clear however that spirituality is not synonymous with religion, and that the assessment of spiritual needs is much more than a tick-box question about religious affiliation. Similarly, spiritual care needs will change as PD progresses, and will both reflect and influence symptoms in other care domains.

It is important that all HCW should be able to offer spiritual care that reaches out to an individual in their time of need. A key aspect of spiritual care involves listening to individual narratives to learn their ‘life-story’. An understanding of what was important to the individual before cognitive decline may help. However, many HCW will not feel equipped to fully explore and address spiritual care needs with their patients. Therefore, referral to SPC and / or chaplaincy services is important where there are unmet spiritual needs. Contact with an appropriate supportive network may also help the family. In Ireland, specialist care may be provided by personnel from various religious denominations, or by lay chaplains, who encompass the beliefs and views of all denominations. Also refer to the Health Services Executive (HSE) Health Services Intercultural Guide<sup>82</sup>.

People who have religious beliefs are often greatly helped by religious support during their illness. Religious beliefs may also become more important to the relatives as death becomes inevitable. Cultural and religious differences around the end of life deserve respect. It is important to recognise the increasing diversity of religious beliefs in Ireland, and to facilitate people of all religions, and those with no organised religion.

Aids to prayer can be helpful when words are not possible, e.g. rosary beads, prayer tablet, or candles. The same applies to physical contact other than just within care tasks, such as hand holding or a gentle massage. This may not be appropriate / acceptable for everyone depending on cultural beliefs.

Where appropriate, the individual and family should be asked about care of the person’s body after death, including religious practices<sup>9</sup>.





# Planning Future Care and End of life Care in PD

## **Recognising advanced disease and limited life expectancy**

Identifying when someone with PD may be approaching their last months of life is very important, e.g. for advance care planning. However, this can be particularly difficult in PD where the disease trajectory typically follows a long progressive decline over many years. Indicators that could suggest that life expectancy is now very limited are often either not recognised or ignored. There is a need for greater openness and discussion of dying and death to address this issue.

Below are some specific indicators that have been suggested to identify that the person with PD has a limited life expectancy (i.e. less than six months; Gold Standards Framework). However, regular assessments are vital to identify the various triggers that may suggest there is a relevant deterioration in a person's condition. Please refer also to the National Clinical Programme for Palliative Care's Needs Assessment Guidance: <http://www.hse.ie/eng/about/Who/clinical/natclinprog/palliativecareprogramme/Resources/needs%20assessment%20guidance.html>.

In some cases symptoms may be reversible, for example where there is an inter-current infection, and it is important to identify and treat any such problems.

Gold Standards Framework - Clinical prognostic indicator<sup>83</sup>:

*Two or more of:*

- Drug treatment not effective or complex regime needed
- More unpredictability of control
- Reduced independence
- Dyskinesias / mobility problems / falls
- Swallowing difficulties
- Psychiatric symptoms

## **Recognising end of life or “actively dying phase”**

It is similarly important to recognise when death is imminent. This allows a change in the focus of care, and gives family members some warning of the approaching death. Recognising imminent death can be particularly challenging in PD where a person has slowly deteriorated over a prolonged period of time. The experience of the guidelines advisory group is also that the dying phase of PD can be unpredictable, sometimes occurring over days or weeks, rather than hour or days.



Staff caring for the person and family members should be advised of this, to avoid carer exhaustion.

### **Withdrawing / rationalising treatment**

**Idiopathic PD.** In the later stages of PD there may be the need to review, and reduce or withdraw, dopaminergic drugs due to lack of drug efficacy and increasing sensitivity to unwanted effects such as visual hallucinations and excessive somnolence. The person with PD and their carers at this stage will sometimes want to reduce medications, exchanging greater levels of physical disability for increased mental clarity (NICE<sup>11</sup> guideline). However some psychological symptoms (e.g. “off” related anxiety) could be made worse, so constant review is required. As a general guide, medication withdrawal should be managed with help from the specialist clinician and Parkinson’s disease Nurse Specialist. Where possible, drug withdrawal should be gradual in order to achieve the best balance between relief of symptoms and minimal side effects.

This situation should however be reviewed on an ongoing basis as frequent adjustments may be required to maintain this balance.

In people who have undergone deep brain stimulation (DBS) surgery for PD, it is recommended that the DBS is only switched off at end of life if it is causing complications for the person, as switching it off may precipitate rigidity / dystonia / pain.

**Parkinsonian Syndromes.** People with MSA and PSP are less responsive to dopamine replacement

therapy, and the benefits of continued use need to be particularly considered in these people as the disease progresses. Timely and regular review and management of the increasing symptom load, addressing physical, psychological, social, and spiritual aspects, is vital to improving quality of life.

### **Preferred place of death**

Part of care planning for people in their home (whether in the community or residential care) is to establish their preferred place of death, bearing in mind that people’s wishes may change over time. One important focus of care at end of life is to avoid an inappropriate hospital admission. However, there is no one setting of care that is “better” than others as a place of death and the critical factor is that the setting can meet the complexity of the person’s care needs at end of life. Appropriate reliable support must be readily accessible to professional and non-professional carers at all times, including weekends and out-of-hours.

### **Support for professionals**

Caring for a person with PD can be challenging at times and at its core it requires MDT input, with knowledge sharing in a timely fashion. Staff also need the basic resources to provide the level of care that they feel the person needs; otherwise their role can be stressful, when they identify a care need but can’t address it.

End of life care can also be difficult for the staff member. It is important that staff support themselves and each other in this area of work. Staff can be supported through peer support, informal debriefing sessions or counselling as appropriate<sup>B4</sup>.

# Carers

An estimated 12,000 people in Ireland are diagnosed with PD<sup>19</sup>, and therefore carers of people with PD represent a sizeable portion of the 187,112 people who were identified as carers in the 2011 census<sup>85</sup>.

While it is important to recognise the relationship of the patient and carer, equally the carer must also be recognised as an individual.

People caring for someone with PD may face many challenges. They may feel like they have already 'lost the person' and experience episodes of grieving. Family may not see the end coming. Carers may be separated from their loved one if they transition to residential care. Carers who experience a high care burden for a long time may experience feelings of emptiness and guilt when the person with PD enters residential care or dies. Carers often have their own support needs. Everyday contact with the outside world might be limited, compounding isolation. Carers should be directed to agencies which can provide invaluable support, such as:

- Family Carers Ireland
- The AIHPC Palliative Hub
- the Parkinson's Association of Ireland
- PSPA Ireland (Supports people with PSP and CBD)
- Multiple System Atrophy Trust
- Move4Parkinson's

Carers should also be offered individual support, as in some cases group support can actually increase carer anxiety (for example if they see people with more advanced disease than their loved one).

Particular challenges to carers of people with PD are the cognitive, personality, and behavioural changes that may occur, especially in advanced stages of disease. These changes are associated with much greater carer burden. Referral to a psychologist may help carers understand and cope.

The Parkinson's Association of Ireland has branches across Ireland which offer support groups to people with PD and their carers. Further information can be found at: <http://www.parkinsons.ie/branches>

**Bereavement care.** Bereavement care is appropriate throughout the disease process, not just at end of life. However, it comes into focus in the end of life phase, during death, and following the relative's death. People with neurological disease and carers are often very frightened of the dying process, and may be more fearful of the process than of the death itself.

If they ask what to expect near the end of life, appropriate information on further support is essential as well as opportunities to share their concerns and fears. Some individuals and their carers will have been told that their condition is not one people die from and therefore may be unprepared, especially for severe progression and rapid death.

Sometimes families need help to recognise the signs that death is approaching. Equally, carers could be the first to recognise and interpret subtle changes in reduced energy, engagement and mood as the neurological condition worsens. Carers should be kept informed about significant changes in care management, for example in oral feeding and hydration.

If there are unmet social, psychological, physical, or spiritual needs, swift referral to SPC and / or chaplaincy services may be required. This is important to prevent severe distress amongst carers.

Bereaved families may benefit from three levels of support ([www.bereaved.ie](http://www.bereaved.ie)<sup>86</sup>): 1. General support and information (e.g. information on the grieving process, help with practical tasks, social support), 2. Extra support (e.g. voluntary bereavement support services, faith groups, community groups), 3. Therapy support (e.g. psychologists, counsellors, doctors).

# Legal issues

There are a number of legal issues which an individual with PD and their families need to address before the end of life phase and before cognitive decline. Often just knowing that the future has been thought about and planned for can bring a sense of calm and relief for everyone involved, making it easier for all to cope. HCW may need to actively prompt a patient to begin discussions about legal and financial affairs.

HCW should be able to advise individuals on the following, and refer on to other supports where appropriate. It is most important that the person with PD has the opportunity to complete any necessary legal documents while they still have the capacity to do this (with a decision making assistant or a co-decision maker, as needed), and so HCW must be proactive in discussing future planning with the patient early in the disease course. Physicians caring for people with PD and related syndromes should be trained and competent in assessing decision making capacity, and all HCW should be familiar with assisted decision making as per the Assisted Decision Making (Capacity) Act 2015<sup>87</sup>. The detailed legislation surrounding assisted decision making is outside the scope of these guidelines.

**Wills.** Everyone aged 18 or over can - and should - make a will to ensure that when they die their property, including a house or flat, money and personal belongings, is distributed exactly as they would wish. Most solicitors in general practice can draw up a simple will. If a will was written long ago, it should be reviewed and updated if necessary.

**Enduring power of attorney.** If a person anticipates that they will become unable to manage his or her legal and financial affairs, he or she may need to appoint someone he or she trusts to act on his or her behalf, such as a relative, friend or solicitor. This legal document can allow another person to make financial and personal care decisions (e.g. where you live) for someone who has lost the ability to make his or her own decisions. The person who makes an enduring power order can

change or revoke it at any time while he or she retains mental capacity; the enduring power of attorney is only registered (comes into force) when the person has lost their decision making capacity.

**Advance care directives.** Sometimes called “living wills”, these allow the person to give instructions about medical treatments that he/she does or doesn’t want if he/she becomes terminally ill and is unable to express his/her wishes. The Assisted Decision-Making (Capacity) Act 2015<sup>87</sup> contains the current legislation on advance care directives, and HCWs caring for a person with PD should become familiar with these. It is anticipated that there will be a specific code of practice developed to guide HCWs in this area.

It is important to note that a person has the right to refuse a particular treatment, but does not have the right to demand specific treatment. For the most updated position, please refer to: [http://www.citizensinformation.ie/en/health/legal\\_matters\\_and\\_health/advance\\_care\\_directives.html](http://www.citizensinformation.ie/en/health/legal_matters_and_health/advance_care_directives.html).

## Brain bank and research

Researchers, including those who are working towards a cure for Parkinson’s disease, are collecting tissue from people with PD following their death. Some people may find it comforting to know that they can contribute to this cause following their death, but this needs to be a very sensitive discussion. Further information about the Dublin Brain Bank can be found at <http://www.rcsi.ie/index.jsp?a=960&n=797&p=331>



# Appendix I

## List of abbreviations

AAN	American Academy of Neurology
ACP	Advance Care Plan
AIHPC	All Ireland Institute of Hospice and Palliative Care
CBD	Corticobasal Degeneration
ECG	Electrocardiogram
EDPA	European Parkinson's disease Association
EFNS	European Federation of Neurological Societies (EFNS)
FDA	U.S Food and Drug Administration
GPP	Good Practice Point
HCW	Healthcare worker
HSE	Health Service Executive
IHF	Irish Hospice Foundation
LBD	Lewy body dementia
MAO-B	Monoamine oxidase B
MDT	Multidisciplinary Team
MSA	Multiple System Atrophy
NA	Neurological Alliance (UK)
NACPC	National Advisory Committee on Palliative Care
NCEC	National Clinical Effectiveness Committee
NCPC	The National Council for Palliative Care (UK)
NELC	National End of Life Care Programme (UK)
NG tube	Nasogastric tube
NHS	National Health Service
NICE	National Institute <i>for</i> Health and Care Excellence
PAI	Parkinson's Association of Ireland
PD	Parkinson's disease
PDD	Parkinson's disease Dementia
PDNS	Parkinson's disease Nurse Specialist
PEG	<i>Percutaneous Endoscopic Gastrostomy</i>
PRN	<i>"pro re nata"</i> i.e. Latin phrase meaning 'use as necessary'.
PSP	Progressive Supranuclear Palsy
PSPA	Progressive Supranuclear Palsy Association
REM	Rapid eye movement
SIGN	Scottish Intercollegiate Guidelines Network
SPC	Specialist Palliative Care
SSRI	Selective Serotonin Reuptake Inhibitor
WHO	World Health Organisation

# Appendix II

## List of steering group and advisory committee members

### STEERING GROUP

Name	Title	Location	Representing
Dr Suzanne Timmons (Chair)	Consultant Geriatrician	Movement Disorder Clinic, St Finbarr's Hospital, Cork	Irish Society of Physicians in Geriatric Medicine (ISPGM)
Dr Siobhan Fox	Project Co-ordinator	Centre for Gerontology and Rehabilitation, University College Cork	
Ms Alison Cashel	PD Nurse Specialist	Dublin	Parkinson's Association of Ireland (PAI)
Prof George Kernohan	Professor of Health Research	University of Ulster, Belfast	
Ms Marie Lynch	Programme Manager	Dublin	Irish Hospice Foundation (IHF)
Dr Ciara McGlade	Consultant Geriatrician	Centre for Gerontology and Rehabilitation, University College Cork	
Dr Tony O'Brien	Consultant in Palliative Medicine	Marymount University Hospice and Hospital, Cork	
Dr Sean O'Sullivan	Consultant Neurologist	Movement Disorders Clinic, Cork University Hospital	Irish Institute of Clinical Neuroscience (IICN)

### ADVISORY GROUP

Name	Title	Location	Representing
Mr Patrick Browne	PD Nurse Specialist,	Galway University College Hospital	
Dr Brian Creedon	Consultant in Palliative Medicine	University Hospital Waterford	
Ms Caroline Dooley Martyn	Director PSPA	Dublin	Progressive Supranuclear Palsy Association Ireland (PSPA)
Ms Mary J Foley	Advanced Nurse Practitioner	Cork	All Ireland Gerontological Nurses Association (AIGNA)
Mr David Hegarty	Physiotherapist,	St Francis Hospice Dublin	Irish Society of Chartered Physiotherapists (ISCP [CPNG branch])

Dr Graham Hughes	Consultant Physician,	St Vincent's University Hospital Dublin	National Clinical Programme Older People  Royal College of Physicians Ireland (RCPI)
Dr Edward Jones	Consultant Geriatrician, Department for Elderly Medicine, Scarborough General Hospital, NHS	Scarborough PD Specialist Palliative Care service, UK	
Prof Tim Lynch	Consultant Neurologist	Mater hospital Dublin	Clinical Lead for National Clinical Programme in Neurology
Mr Brian Magennis	PD Nurse Specialist	Mater hospital Dublin	
Ms Elaine O'Connor	Senior Occupational Therapist	Dublin	Irish Gerontological Society (IGS)
Mr Diarmaid O'Sullivan	Campaigns / Research Manager	Dublin	Family Carers Ireland
Ms Catryn Power	Person with PD	Cork	Parkinson's Association of Ireland (PAI)
Ms Margaret Richardson	PD Nurse Specialist	Mid-Western Regional Hospital Limerick	
Ms Katie Rigg	MSA Nurse Specialist	London	MSA Trust
Ms Sheighle Sheridan	Senior Medical Social Worker	Mater Hospital Dublin	Irish Association of Social Workers (IASW)
Dr Catherine Sweeney	GP, Medical Director, Services for Older People	Marymount University Hospice and Hospital Cork	All Ireland Institute of Hospice and Palliative Care (AIHPC)  Irish College of General Practitioners (ICGP)
Ms Jean Barber	CNS Palliative Care	St. Michael's Hospital, Dun Laoghaire, Dublin	Irish Association for Palliative Care (IAPC)
Dr Janis Miyaski	Clinical Neurologist and Ass. Prof. Dept. of Medicine, University of Alberta	Integrated Palliative Care/ Movement Disorders clinic, Alberta, Canada	
Ms Margaret Mullarney	Director, Move 4 Parkinsons and Person with PD	Dublin	Move 4 Parkinsons
Ms Heather Coetzee	Speech Therapy Manager	Dublin	Health and Social Care Professions Directorate
Dr Denise Hayes	SpR in Palliative Medicine,	Waterford University Hospital	
Ms Deirdre Shanagher	Development officer	Irish Hospice Foundation, Dublin	



# Appendix III

## Grading criteria used by existing guidelines

### NICE – Classification of recommendations and evidence statements

Grade	Evidence	Type of Evidence
A	<ul style="list-style-type: none"> <li>• At least one meta-analysis, systematic review, or randomised controlled trial (RCT) that is rated as 1++, and is directly applicable to the target population, or</li> <li>• A systematic review of RCTs or a body of evidence that consists principally of studies rated as 1+, is directly applicable to the target population and demonstrates overall consistency of results, or</li> <li>• Evidence drawn from a NICE technology appraisal</li> </ul>	1++ High-quality meta-analysis (MA), systematic reviews (SR) of randomized controlled trials (RCTs), or RCTs with a very low risk of bias 1+ Well-conducted MA, SR or RCTs, or RCTs with a low risk of bias
B	<ul style="list-style-type: none"> <li>• A body of evidence that includes studies rated as 2++, is directly applicable to the target population and demonstrates overall consistency of results, or</li> <li>• Extrapolated evidence from studies rated as 1++ or 1+</li> </ul>	2++ High-quality SR of case-control or cohort studies 2++ High-quality case-control or cohort studies with a very low risk of confounding, bias or chance and a high probability that the relationship is causal 2+ Well-conducted case-control or cohort studies with a low risk of confounding, bias or chance and a moderate probability that the relationship is causal
C	<ul style="list-style-type: none"> <li>• A body of evidence that includes studies rated as 2+, is directly applicable to the target population and demonstrates overall consistency of results, or</li> <li>• Extrapolated evidence from studies rated as 2++</li> </ul>	
D	<ul style="list-style-type: none"> <li>• Evidence level 3 or 4, or</li> <li>• Extrapolated evidence from studies rated as 2+, or</li> <li>• Formal consensus</li> </ul>	3 Non-analytic studies (for example case reports, case series) 4 Expert opinion, formal consensus
D(GPP)	A good practice point (GPP) is a recommendation for best practice based on the experience of the Guideline Development Group	

## SIGN - Classification of recommendations and evidence statements

LEVELS OF EVIDENCE	
1++	High quality meta-analyses, systematic reviews of RCTs, or RCTs with a very low risk of bias
1+	Well conducted meta-analyses, systematic reviews, or RCTs with a low risk of bias
1-	Meta-analyses, systematic reviews, or RCTs with a high risk of bias
2++	High quality systematic reviews of case control or cohort studies
2+	Well conducted case control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal
2-	Case control or cohort studies with a high risk of confounding or bias and a significant risk that the relationship is not Causal
3	Non-analytic studies, e.g. case reports, case series
4	Expert opinion
GRADES OF RECOMMENDATION	
A	At least one meta-analysis, systematic review, or RCT rated as 1++, and directly applicable to the target population; or A body of evidence consisting principally of studies rated as 1+, directly applicable to the target population, and demonstrating overall consistency of results
B	A body of evidence including studies rated as 2++, directly applicable to the target population, and demonstrating overall consistency of results; <i>or</i> Extrapolated evidence from studies rated as 1++ or 1+
C	A body of evidence including studies rated as 2+, directly applicable to the target population and demonstrating overall consistency of results; <i>or</i> Extrapolated evidence from studies rated as 2++
D	Evidence level 3 or 4; <i>or</i> Extrapolated evidence from studies rated as 2+
GOOD PRACTICE POINTS (GPP)	Recommended best practice based on the clinical experience of the guideline development group.

## American Academy of Neurology – Classification of Evidence

Recommendation grade	Evidence
A	<ul style="list-style-type: none"> <li>Established as effective, ineffective, or harmful for the given condition in the specified population.</li> <li>Level A rating requires at least two consistent Class I studies</li> </ul>
B	<ul style="list-style-type: none"> <li>Probably effective, ineffective, or harmful for the given condition in the specified population.</li> <li>Level B rating requires at least one Class I study or at least two consistent Class II studies.</li> </ul>
C	<ul style="list-style-type: none"> <li>Possibly effective, ineffective, or harmful for the given condition in the specified population.</li> <li>Level C rating requires at least one Class II study or two consistent Class III studies.</li> </ul>
U	<ul style="list-style-type: none"> <li>Data inadequate or conflicting given current knowledge, treatment is unproven</li> </ul>

## Movement Disorder Society – Classification of Evidence

Efficacy conclusions	Definition	Required evidence
Efficacious	Evidence shows that intervention has a positive effect on studied outcomes	Supported by data from at least 1 high-quality (score $\geq$ 75%) RCT without conflicting level-I data
Likely efficacious	Evidence suggests, but is not sufficient to show, that the intervention has a positive effect on studied outcomes	Supported by data from any Level-I trial without conflicting level-I data
Unlikely efficacious	Evidence suggests that the intervention does not have a positive effect on studied outcomes	Supported by data from any Level-I trial without conflicting level-I data
Non-efficacious	Evidence shows that the intervention does not have a positive side effect on studied outcomes	Supported by data from at least 1 high-quality (score $\geq$ 75%) RCT without conflicting level-I data.
Insufficient evidence	There is not enough evidence either for or against efficacy of the intervention in treatment of Parkinson's disease	All the circumstances not covered by the previous statements

## European Federation of Neurological Societies - Classification of Evidence

Recommendation grade	Evidence
A	established as effective, ineffective, or harmful, requires at least one convincing class I study or at least two consistent, convincing class II studies
B	probably effective, ineffective, or harmful, requires at least one convincing class II study or overwhelming class III evidence
C	possibly effective, ineffective, or harmful rating, requires at least two convincing class III studies
GPP	where there is insufficient scientific evidence, a consensus statement (“good practice point”)

## Simplified grading scheme used in the Canadian guidelines

Recommendation grade	Evidence
A	Established as effective, ineffective, or harmful for the given condition in the specified population.
B	Probably effective, ineffective, or harmful for the given condition in the specified population.
C	Possibly effective, ineffective, or harmful for the given condition in the specified population.
D	Expert opinion, formal consensus.
U	Data inadequate or conflicting given current knowledge, treatment is unproven.
GPP	Good practice point.



# Appendix IV

## Search strategy used for systematic review

As part of the systematic review, four databases were searched in February and March 2015. The databases searched were: Emabase, Medline, Cinahl, and PsychInfo. The review included articles published since January 2010. The search terms used, limiters applied, and number of 'hits' are reported in the tables below.

### MEDLINE (via Ovid) searched on 27/02/15

	Search terms	Hits
1	exp Parkinson disease/	48907
2	parkinson disease.mp.	54162
3	Parkinson*.mp.	83216
4	exp Supranuclear palsy, progressive/	1922
5	exp Lewy Body Disease/	2204
6	exp Multiple System Atrophy/	2714
7	exp Basal Ganglia Disease/	86771
8	exp Parkinsonian Disorders/	59637
9	1 or 2 or 3 or 4 or 5 or 6 or 7 or 8	110466
10	exp Therapeutics/	3416475
11	exp Pharmaceutical Preparations/	620047
12	Palliative Care/	41286
13	Crisis Intervention/ or Intervention Studies/ or Early Medical Intervention/	13548
14	therapy.mp.	1653227
15	drug*.mp.	2079695
16	intervention.mp.	331006
17	evaluation.mp.	1152185
18	exp Qualitative Research/	22139
19	10 or 11 or 12 or 13 or 14 or 15 or 16 or 17 or 18	6463589
20	exp Muscle Rigidity/	1789
21	exp Pain/	314310
22	Dysphagia.mp. or exp Deglutition Disorders/	49293
23	exp Dyspnea/	15978
24	exp Nausea/	16021
25	Pressure Sores.mp. or exp Pressure Ulcer/	10384
26	exp Fatigue/	21107
27	exp Sleep Disorders/	63521
28	exp "Disorders of Excessive Somnolence"/	5575
29	exp Hypotension, Orthostatic/	4941
30	exp Memory/ or exp Memory Disorders/	116962
31	exp Cognition/	112909
32	exp Hallucinations/	9173
33	exp Depression/	79319
34	exp Anxiety/	57586
35	exp Delirium/	6027
36	exp Accidental Falls/	16113
37	20 or 21 or 22 or 23 or 24 or 25 or 26 or 27 or 28 or 29 or 30 or 31 or 32 or 33 or 34 or 35 or 36	812549
38	9 and 19 and 37	4201
39	limit 37 to humans; yr="2010 -Current"	1128

### Cinahl (via Ebsco) searched on 12/03/15

	Search terms	Hits
S1	(MM "Parkinson Disease") OR (MM "Lewy Body Disease") OR (MM "Supranuclear Palsy, Progressive") OR "multiple system atrophy" OR (MM "Parkinsonian Disorders") OR "corticobasal degeneration"	11,356
S2	"treatment" OR (MM "Treatment Outcomes+") OR (MM "Drugs+") OR (MM "Palliative Care") OR (MM "Terminal Care+") OR (MH "Intervention Trials") OR (MM "Nursing Interventions") OR (MM "Crisis Intervention") OR (MM "Experimental Studies+") OR (MM "Evaluation") OR (MM "Drug Evaluation+") OR (MM "Program Evaluation") OR (MM "Evaluation Research") OR (MM "Quality of Life+") OR (MM "Health and Life Quality (Iowa NOC) (Non-Cinahl)") OR (MM "Qualitative Studies+")	617,783
S3	(MM "Pain+") OR "rigidity" OR (MM "Swallowing Therapy") OR "dysphagia" OR (MM "Dyspnea+") OR (MM "Nausea") OR (MM "Nausea Care (Saba CCC)") OR (MM "Nausea and Vomiting+") OR (MM "Pressure Ulcer+") OR (MH "Fatigue") OR (MH "Sleep Disorders") OR (MM "Hypotension, Orthostatic+") OR (MM "Memory Disorders+") OR (MM "Cognition Disorders+") OR (MM "Depression+") OR (MM "Anxiety+") OR (MM "Hallucinations+") OR (MM "Delirium") OR (MM "Accidental Falls")	201,226
S4	S1 AND S2 AND S3	372
S5	S1 AND S2 AND S3 Limit: humans Limit Jan 2010 to Mar 2015	115

### PsychInfo (via Ebsco) searched on 12/03/15

	Search terms	Hits
S1	SU ( 'Parkinson* disease' OR 'Parkinsonian' OR 'Parkinson*' ) OR SU 'progressive supranuclear palsy' OR SU ( 'lewy body dementia' OR 'Lewy Body disease ) OR SU 'multiple system atrophy' OR SU 'parkinson disease dementia' OR SU 'corticobasal degeneration'	19,069
S2	SU therapy OR SU drug* OR SU intervention OR SU evaluation study OR SU ( 'qualitative study' OR 'qualitative research' ) OR SU pharmatherapy OR SU pharmacological OR SU ( 'palliative therapy OR 'palliative care' )	477,552
S3	SU 'muscle rigidity' OR SU pain OR SU ( 'dysphagia' OR 'swallow difficulty' ) OR SU ( dyspnea OR 'breathing difficulty' OR 'shortness of breath' ) OR SU nausea OR SU ( 'decubitus' OR 'pressure sore' OR 'pressure ulcer' ) OR SU fatigue OR SU 'sleep disorder' OR SU ( 'orthostatic hypotension' OR 'postural hypotension' ) OR SU ( 'memory disorder' OR 'cognition' ) OR SU ( 'depression' OR 'delirium' OR 'anxiety' ) OR SU 'visual hallucination' OR SU (falls OR fall OR balance)	297,295
S4	S1 AND S2 AND S3	611
S5	S1 AND S2 AND S3 <b>Limiters</b> - Publication Year: 2010-2015; Published Date: 20100101-20150331; Population Group: Human	204

### Embase (via Elsevier) searched on 13/03/15

1	'parkinson disease'/exp OR 'parkinson disease'	101,782
2	'parkinsonian'	13,096
3	'progressive supranuclear palsy'/exp OR 'progressive supranuclear palsy'	4,942
4	'diffuse lewy body disease'/exp OR 'diffuse lewy body disease'	4,574

5	'multiple system atrophy'/exp OR 'multiple system atrophy' OR 'shy drager syndrome'/exp OR 'shy drager syndrome'	5,036
6	'parkinson disease dementia'	144
7	'corticobasal degeneration'/exp OR 'corticobasal degeneration'	2,198
8	1 OR 2 OR 3 OR 4 OR 5 OR 6 OR 7	113,272
9	'therapy'/exp OR 'therapy'	8,948,294
10	'intervention studies'/exp OR 'intervention studies'	29,439
11	'drug mechanism'/exp OR 'drug mechanism'	275,004
12	palliative AND ('therapy'/exp OR therapy)	96,122
13	'evaluation research'/exp OR 'evaluation research' OR 'evaluation study'/exp OR 'evaluation study'	194,283
14	'qualitative analysis'/exp OR 'qualitative analysis' OR 'qualitative research'/exp OR 'qualitative research'	75,013
15	9 OR 10 OR 11 OR 12 OR 13 OR 14	9,233,762
16	'muscle rigidity'/exp OR 'muscle rigidity'	8,850
17	'pain'/exp OR 'pain'	1,095,915
18	'dysphagia'/exp OR 'dysphagia'	51,627
19	'dyspnea'/exp OR 'dyspnea'	108,523
20	'nausea'/exp OR 'nausea'	197,539
21	'decubitus'/exp OR 'decubitus'	18,766
22	'fatigue'/exp OR fatigue	167,846
23	'sleep disorder'/exp OR 'sleep disorder'	169,471
24	'orthostatic hypotension'/exp OR 'orthostatic hypotension'	17,887
25	'memory disorder'/exp OR 'memory disorder'	54,425
26	'cognition disorder'/exp OR 'cognition disorder' OR 'cognitive defect'/exp OR 'cognitive defect'	103,578
27	'visual hallucination'/exp OR 'visual hallucination'	4,948
28	'depression'/exp OR 'depression'	494,142
29	'anxiety'/exp OR 'anxiety'	236,805
30	'falling'/exp OR 'falling' OR 'falls'	92,104
31	'delirium'/exp OR 'delirium'	21,691
33	16 OR 17 OR 18 OR 19 OR 20 OR 21 OR 22 OR 23 OR 24 OR 25 OR 26 OR 27 OR 28 OR 29 OR 30 OR 31	2,183,256
34	8 AND 15 AND 33	19,145
	#34 AND [humans]/lim AND [2010-2015]/py	7,600

# Appendix V

## Submissions received during public consultation

A total of 21 submissions were received. Personal submissions included submissions from allied health professionals, family carers of people with a Parkinsonian syndrome, and others who expressed a personal interest in the area. The organisations which had members provide feedback are listed below. Note that the guidelines were also distributed to organisations in addition to those listed below, however only those listed provided feedback.

Two international experts on palliative care for people with neurodegenerative diseases were invited to provide feedback as part of this process, prior to the wider public consultation. These experts were Professor David Oliver, Consultant in Palliative Medicine, Rochester, and Honorary Reader, the University of Kent; and Professor Anette Schrag, Professor of Clinical Neurosciences, University College London.

- All Ireland Gerontological Nurses Association (AIGNA)
- Association of Occupational Therapists of Ireland (AOTI) - Palliative Care and Oncology Advisory Group
- Health Service Executive Quality Improvement Division
- Irish Association of Speech and Language Therapists (IASLT)
- Irish College of General Practitioners (ICGP) - Quality in Practice Committee
- Irish Institute of Clinical Neuroscience (IICN)
- Irish Society of Chartered Physiotherapists (ISCP) - Chartered Physiotherapists in Neurology and Gerontology Clinical Interest Group
- Irish Society of Physicians in Geriatric Medicine (ISPGM)
- Multiple System Atrophy Trust
- National Clinical Programmes for Neurology, Palliative Care, and Older Persons
- The National Social Work Organisation of Ireland - Head Medical Social Workers Group and the Social Workers in Aging Special Interest Group
- Nursing Homes Ireland (NHI)

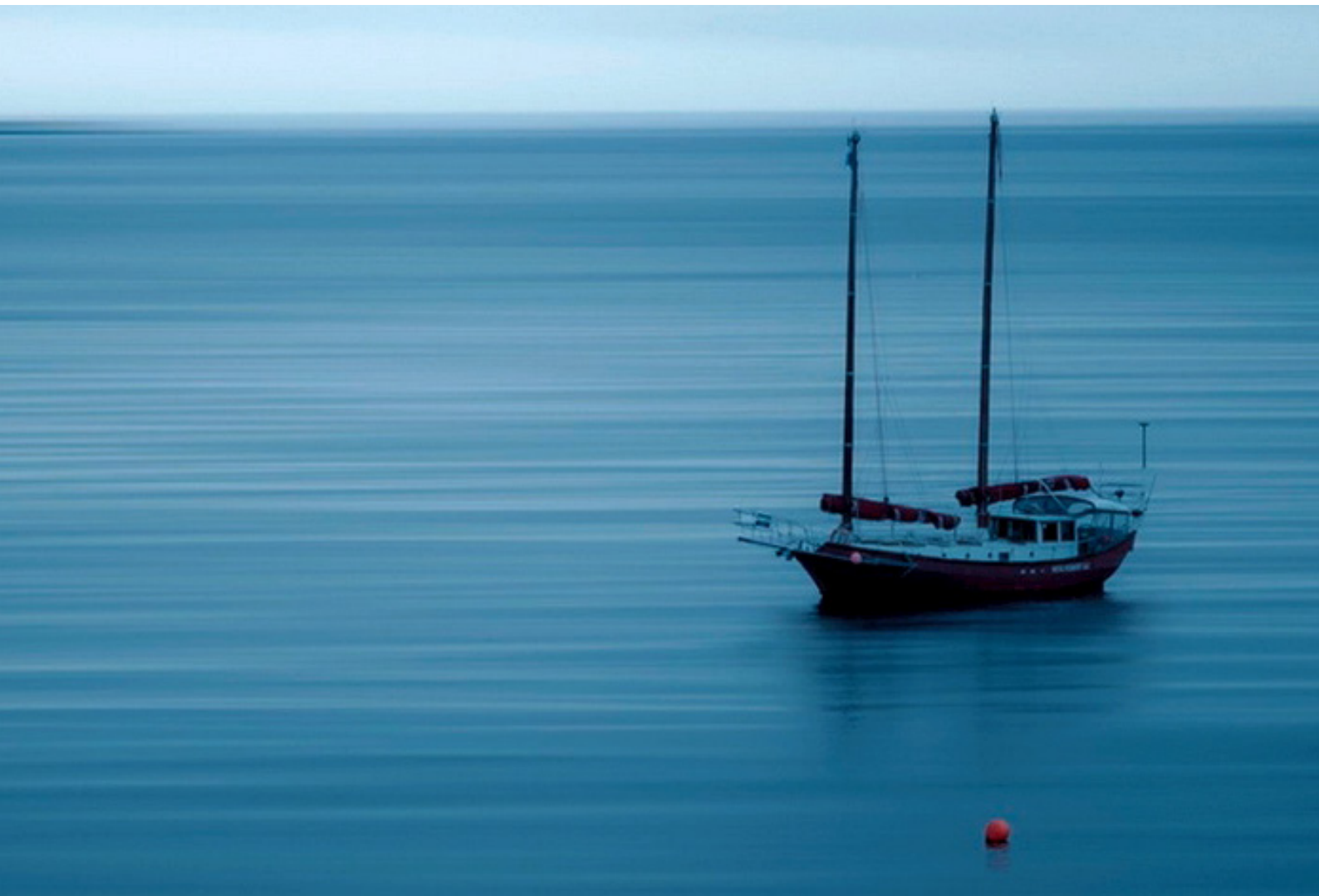


# Appendix VI

## Multidisciplinary Team Workers

At the time of diagnosis the professionals involved in the care of the person with PD may be the GP, consultant neurologist / geriatrician with a speciality in Parkinson's disease, or Clinical Nurse Specialist in PD. However throughout the disease an increasing number of professionals may become involved, including, but not exclusive to:

- Continenence services
- Dietitians
- Neuro / Clinical psychologists
- Occupational therapists
- Pastoral or spiritual care
- Pharmacy
- Physiotherapists
- Professional carers
- Psychiatry
- Public health nurses
- Rehabilitation physicians
- Social services managers
- Social workers
- Specialist palliative care
- Speech and language therapists
- Voluntary organisations, e.g. the Parkinson's Association of Ireland, PSPA Ireland, MSA Trust



# Appendix VII

## List of relevant websites

The website of the **Parkinson's Association of Ireland (PAI)** the national support and advocacy charity for Parkinson's in Ireland. Information for professional healthcare workers is available on the website, including the following link:

[http://www.parkinsons.ie/professionals\\_palliativecare](http://www.parkinsons.ie/professionals_palliativecare)

The **Multiple System Atrophy Trust** is a UK based charity supporting people affected by multiple system atrophy. They also have information about support for healthcare professionals at the following link:

<http://www.msatrust.org.uk>

**PSPA Ireland** is a voluntary organisation which supports people who have or are affected by Progressive Supranuclear Palsy or Corticobasal Degeneration. Information on both PSP and CBD is available through their website:

[www.pspaireland.ie](http://www.pspaireland.ie)

The **Irish Hospice Foundation (IHF)** is a national charity dedicated to all matters relating to dying, death and bereavement in Ireland. The IHF website provides a wealth of information, including that specific to the care of people with advancing neurological illness, available at:

<http://hospicefoundation.ie/healthcare-programmes/palliative-care/palliative-care-for-all/advancing-neurological-illness/13398-2/>

The website **'Bereaved.ie'** has links to a range of supports and training for people who work (in a voluntary capacity or as a professional) with people who have been bereaved.

[http://hospicefoundation.ie/bereavement/?gclid=CJGWgu-mp8ICFQPhtAodtEIA\\_Q](http://hospicefoundation.ie/bereavement/?gclid=CJGWgu-mp8ICFQPhtAodtEIA_Q)

Further information about the **Dublin Brain Bank** tissue donation can be found at: <http://www.rcsi.ie/index.jsp?a=960&n=797&p=331>

The websites for the Health Service Executive **National Clinical Programmes** contain useful information, including downloadable documents and links to other websites/resources.

The National Clinical Programme for Palliative care website is available at:

<http://www.hse.ie/eng/about/Who/clinical/natclinprog/palliativecareprogramme/>

The National Clinical Programme for Neurology website is available at:

<http://www.hse.ie/eng/about/Who/clinical/natclinprog/neurology/>

The National Clinical Programme for Older People website is available at:

<http://www.hse.ie/eng/about/Who/clinical/natclinprog/olderpeopleprogramme/>

**All Ireland Institute of Hospice and Palliative Care (AIHPC)**, with the palliative care sector, have developed 'The Palliative Hub' to act as a gateway to information and resources about palliative care on the island of Ireland. There are sub-sections of the Hub aimed at different groups, as follows:

The Palliative Hub – Adult is aimed at adults with palliative care needs, their family members and carers, including the wider public engaged with someone with a palliative need:

<http://aiihpc.org/palliative-hub/adult-public/>

The Palliative Hub – Learning Platform provides an online learning environment for both professionals and the public to learn about palliative care and build upon their current knowledge:

<http://aiihpc.org/palliative-hub/learning-platform/>

The Palliative Hub – Professional is currently in development (launching Spring 2016) and will be aimed at health and social care professionals, academics, researchers, educationalists and volunteers working in the palliative care field or who have a need to learn more about palliative care:

<http://aiihpc.org/palliative-hub/professional-academic-and-research/>

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