Framework for professionals supporting patients deteriorating with Parkinson's Disease (PD)

PROBLEM/STAGE	DESCRIPTION	ACTION
Recognising advancing disease	The disease trajectory of PD typically follows a long progressive decline over many years. Identifying when someone with PD is approaching their last months of life is challenging but important. Below are some specific indicators that have been suggested to recognise advancing disease. Be aware that a person may not experience all of these. • Hallucinations • Weight loss • LRTI/Recurrent Pneumonia • Swallowing difficulties • Increasing number of falls • Drug treatment not effective or a more complex regime is needed. It is important to also note other significant life events that a person is experiencing, for example bereavement.	Identifying when someone with PD is experiencing a period of flux can trigger: • Conversations to help identify priorities and wishes of the patient. • Opportunities to record these informally or formally and share with relevant HCPs (e.g., by completing a ReSPECT form) • Opportunities to create an 'anticipatory care plan'. • Entering the patient's data onto an End-of-Life Care register/EPaCCS
Sudden deterioration of PD symptoms	It is unusual for a rapid deterioration to be caused by PD alone. Ensure a complete assessment has occurred to identify reversible causes. The following should be considered: • Intercurrent illness (e.g., UTI, chest or other infection, aspiration, dehydration, constipation) • Change in medication – consider all medications, not only a change in anti-Parkinson's treatment. For example, has an anti-cholinergic or opiate drug been commenced?	• Clinical assessment to exclude intercurrent illness. • Review medication changes • Establish wishes of the patients regarding ceilings of treatment / investigation / place of care. • Ensure any changes in wishes have been updated on ACP documentation such as ReSPECT form, EPaCCS, Connecting care.

Typical / Anticipated Non motor Symptoms	People with PD report an average of at least eight non motor symptoms (NMS), which are often more difficult to recognise and treat than motor symptoms but cause significant distress. Neuropsychiatric: • Psychosis • Agitation • Anxiety / Depression • Cognitive impairment Autonomic dysfunction: • Bladder • Postural hypotension • Erectile dysfunction • Sialorrhoea Pain GI symptoms: • Constipation • Gastroparesis Speech dysfunction: • Dysarthria • Dysphagia Sleep dysfunction	Consider assessment of NMS using validated tool - such as Fillable NMS questionnaire.pdf (parkinsons.org.uk) • Consider referral to PD service for optimisation of symptom control.
Parkinson's treatment related issues	Dopaminergic medications offer significant benefit for motor symptoms but motor fluctuations and dyskinesias may occur at even moderate stages of the disease.	• Ensure all Parkinson's medications are given at the times suggested and that brand is consistent. • Establish whether swallowing difficulties are contributing to concordance issues with medications.

	Adverse neuropsychiatric and autonomic side effects of these medications maybe prominent and include: • Nausea and vomiting • Confusion / Hallucinations • Dyskinesia • Low blood pressure • Drowsiness • Impulse control disorder (with dopamine agonists) Simplifying medications in advanced PD to focus mostly on Levodopa may be preferable to reduce medication burden and ameliorate side effects, particularly those associated with adjunctive therapies such as Dopamine agonists and anti-cholinergics. Changes in medication should be in consultation with the patient's usual PD team. Be aware that some patients report a reduced efficacy when medication brand is switched (for example switching from Sinemet plus 100/25 to generic Co-Careldopa 100/25). Abrupt cessation or withholding of dopaminergic medications is contraindicated in almost all circumstances due to the risk of worsening motor and non-motor symptoms in the 'off state' and also because of the risk of neuroleptic-like malignant syndrome (L-dopa) and Dopamine Agonist Withdrawal Syndrome (Dopamine Agonists).	• If suspicion that a change in medication has caused someone to feel unwell, seek advice from the PD team. • Be alert to features of neuroleptic-like malignant syndrome in anyone who has stopped treatment abruptly • Changes in PD medication should generally take place in consultation with the patient's usual PD team
Inability to swallow Parkinson's medications	People with PD are at greater risk of their swallowing deteriorating – either in the context of intercurrent illness or as part of the condition.	• Rule out additional pathology if someone presents with swallowing difficulties for the first time.

(when this has arisen acutely)	Avoid the abrupt withdrawal of Parkinson's medications and missed doses. Be cautious in diagnosing dying in a patient with PD who has missed medication doses – the 'off' state may look like dying and be brought about by something like constipation-associated non-absorption of medication.	• Actively look for and treat intercurrent illness (infection, constipation, urinary retention, electrolyte abnormalities, oral candidiasis) • Offer good mouth care. • Do not alter treatment regimens or timings without advice. • **NEVER** prescribe dopamine antagonists (E.g., haloperidol, metoclopramide, prochlorperazine) • Where possible seek specialist input from the PD team early • If someone is struggling to swallow food and fluids, as well as medication, urgent speech and language assessment should be sought. *See hyperlink for flow chart and conversion tables to support maintenance of PD meds in patients with swallowing difficulties (BNSSG guidelines 2018)* https://remedy.bnssg.icb.nhs.uk/media/5765/management-of-community-based-parkinsons-patients-unable-to-swallow-med_v1.pdf **Please note caution in conversions**. If there is a high risk of delirium, consider using the 'Optimal' calculator: http://parkinsonscalculator.com/
Recognising and managing the dying phase	It is important to recognise when someone is 'actively' dying as this allows a change in the focus of care and gives family some warning of approaching death. Recognising imminent death can be particularly challenging in PD, where a person has slowly deteriorated over a prolonged period of time. The dying phase of PD can be unpredictable, sometimes occurring over days or weeks, rather than hours or days.	Establish the wishes of the person and family in relation to their care at the dying phase (including place of care) • Exclude reversible causes. • Ensure no recent discontinuation / omission of usual anti-parkinsonian medications before diagnosing dying. • Continue usual medications if able. *Please see link to BNSSG guidelines 2018:*

	It may be helpful to consider the following: • Exclude reversible causes. • The 'Off' phase with significant rigidity / bradykinesia may be mistaken for dying. A Deep Brain Stimulator (DBS) must be removed before cremation.	https://remedy.bnssg.icb.nhs.uk/media/5765/management-of-community-based-parkinsons-patients-unable-to-swallow-med_v1.pdf If the oral route is no longer possible and you are considering converting to a PD medication patch this should usually be done in liaison with the patient's usual PD team.
Management of common symptoms in the dying phase (last days of life)	Pain • There are a number of different causes of pain in patients with PD: musculoskeletal (including rigidity), dystonic, neuropathic (often under recognised in PD). • The approach to pain should follow the principles of the WHO analgesic ladder, recognising that analgesia may need to be given via the TD/SC route if patients are unable to swallow safely. Please note the increased risk of constipation with opiates. • Remember to consider use of PD medications to optimise management of motor symptoms causing pain. Nausea and vomiting • Consider treating any underlying causes of nausea that remain appropriate. • Metoclopramide, Haloperidol, Cinnarizine, Prochlorperazine should not be used in PD, as they have antagonist effects at dopamine receptors. Levomepromazine and Cyclizine should be used with caution. • Hiccups are an occasional SE of agonists where no other cause identifiable.	To lessen rigidity, ensure dopaminergic medication is given on time according to the individual's usual regimen. If someone is on an apomorphine pump, it may be appropriate to maintain the infusion to prevent rigidity and pain - seek advice from the PD team. If PD medications have been optimised and rigidity remains a problem, it may be appropriate to use midazolam via subcutaneous infusion to manage this alongside other analgesia. Ondansetron (PO/SC) and Domperidone (PO) are the anti-emetics of choice in the management of nausea and vomiting in PD. Cyclizine and other anticholinergics risk worsening of confusion, so should be avoided if possible. If agitation persists, despite optimisation of dopaminergic medication and treatment of reversible causes then consider: • Whether a switch to a Rotigotine patch may have contributed, particularly if patient is elderly or previously agonist naïve. • Use of Benzodiazepines (most likely SC midazolam if not swallowing) • Seek specialist advice if persistent despite this. • Review response to medications and consider further changes. • Consider referral to specialist palliative care team

	Agitation and hallucinations • Consider treating any underlying causes (e.g., constipation, urinary retention, pain) • Dopaminergic medications can contribute towards confusion and hallucinations, so these may need to be adjusted. • Antipsychotic medications have antagonist effects at dopamine receptors so should be avoided	
Dopaminergic medication in last days	Consensus opinion favours continuation of dopaminergic treatment in the last days. Where possible, usual oral medication can be continued. Non-oral dopaminergic therapies (Duodopa, apomorphine) can be continued where regime already established. Where oral route not possible a Rotigotine patch can be used – this is off licence and based on limited case studies. Terminal delirium / agitation is common in people with PD and there are concerns that dopamine agonists (e.g., Rotigotine) may play a role here particularly in patients who are agonist naïve. Until greater evidence is available, we suggest discussion with Parkinson's service (PD nurse and / or consultant) when initiating Rotigotine for the first time in the terminal phase.	Discussion with Parkinson's service (PD nurse and / or consultant) when initiating Rotigotine for the first time in the terminal phase.