# Diagnostic, management and nursing challenges of less common dementias: Parkinsonian dementias and Huntington's disease

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## Abstract

Background: Although most cases of dementia are caused by Alzheimer's disease or vascular dementia, around 10-15% of cases are due to other disorders, including dementias with Parkinsonian features, Huntington's disease, frontotemporal dementia, human immunodeficiency virus (HIV), and alcohol.

Aims: These less common dementias are important as they may have differing clinical features and require different approaches to diagnosis and management. This paper seeks to provide relevant information for nurses about symptoms, diagnosis and management of some of the less common dementias.

Methods: This is one of two connected papers, and provides a clinical overview of Parkinsonian dementias and Huntington's disease. It provides a narrative, rather than systematic, review of the literature.

Findings: Parkinsonian dementias comprise Parkinson's disease dementia, dementia with Lewy bodies and so-called Parkinson's-plus syndromes (multi-system atrophy, progressive supranuclear palsy, and corticobasal degeneration). Huntington's disease is an inherited neuropsychiatric condition. Each has a distinctive clinical picture, with combinations of cognitive, neuropsychiatric and neurological symptoms but approaches to treatment and care are essentially supportive.

Conclusions: Nurses have an essential role in supporting people with dementia, as well their families and carers, throughout the course of dementia from diagnosis to end of life care. They are often best placed and have the necessary skills to create appropriate care plans and to provide care management.

## Introduction

Over 537,000 people in the UK have a dementia diagnosis (Alzheimer's Research UK, 2020). The most common causes of dementia are Alzheimer's disease (AD) (accounting for two-thirds of all cases) and vascular dementia, but there are also many lesser-known causes, which altogether perhaps account for 10-15% of all cases. Therefore, although these disorders are 'less common', some of them are far from rare. The less common forms of dementia differ in aetiology, incidence, clinical features, age of onset, prognosis and ability to treat (Dening and Babu Sandilyan, 2015a; 2015b). These forms of dementia may often cause diagnostic and management difficulties.

Less common dementias often present at younger ages, with wide-ranging signs and symptoms. They may be more likely to be part of an underlying neurological syndrome. Prompt diagnosis enables careful and considered care planning to occur at an early stage.

The diagnosis of dementia mostly remains a clinical process. Investigations, such as neuroimaging, are important in terms of establishing the cause of the dementia, and there is currently much interest in validating biomarkers, especially in AD (Olsson et al, 2016). The heavy emphasis on memory impairment in clinical criteria lends itself more towards a diagnosis of Alzheimer's dementia and is less help in diagnosing less common causes (Dening and Babu Sandilyan, 2015a; 2015b).

This paper is the first of two about some of the important and less common dementias, in which we consider their diagnosis and management. The authors will also discuss the provision of support to patients and families, including the contribution of nursing. Part one includes the various types of dementia with Parkinsonian features and Huntington's disease; Part 2 will discuss frontotemporal dementia, HIV dementia and alcoholic dementia.

## Dementias with Parkinsonian features

This is a mixed group of disorders (see Table 1), including Lewy body spectrum disorders (encompassing Parkinson's disease (PD), Parkinson's disease dementia (PDD), and Lewy body dementia (DLB)), and the so-called Parkinson-plus syndromes, which are other forms of dementia with Parkinsonian features, including corticobasal degeneration (CBD), multi-system atrophy (MSA) and progressive supranuclear palsy (PSP). The Lewy body disorders and MSA are synucleinopathies—that is, they are characterised by accumulation of the protein α-synuclein in the central nervous system. In contrast, PSP and CBD are characterised by accumulation of insoluble tau protein (Armstrong and McFarland, 2019).

**Table 1: Summary of the key features of dementias with Parkinsonian features**

| **Synucleinopathies** | Pathology | Features |
|---|---|---|
| Parkinson's disease dementia | Lewy bodies, cholinergic deficits | ***Likely***: Early motor symptoms, early visuospatial and executive dysfunction, retrieval memory impairment, REM sleep behaviour disorder, visual hallucinations<br><br>***Less likely:*** Early cognitive decline |
| Lewy body dementia | Lewy bodies in the cortex, brainstem and limbic regions, dopamine and acetylcholine abnormalities | ***Likely***: Early falls, retrieval memory problems, early and fluctuating cognitive impairments in attention, visuospatial and executive function, antipsychotic sensitivity, hyposmia, REM sleep behaviour disorder, early visual hallucinations.<br><br>***Less likely***: Autonomic dysfunction |
| Multi-system atrophy | Abnormal proteins causing glial and myelin dysfunction and neurodegeneration. | ***Likely:*** Early autonomic symptoms, cerebellar signs, parkinsonism, restless legs, speech impairment, laryngeal stridor, relatively preserved cognitive function.<br><br>***Less likely***: Hyposmia |
| **Tauopathies** | | |
| Progressive supranuclear palsy | Accumulation of tau in neurofibrillary tangles with degeneration of nigral and subcortical areas | ***Likely***: Early falls, postural instability, gait problems, vertical supranuclear gaze palsy, pseudobulbar palsy, early and severe cognitive impairment (frontal)<br><br>***Less likely:*** REM sleep behaviour disorder, olfactory problems |
| Corticobasal degeneration | Degeneration of the cerebral cortex and deeper brain regions. | ***Likely***: Limb rigidity, bradykinesia, postural instability, alien limb, visuospatial deficits, apraxia, dysarthria and aphasia, episodic memory reasonably preserved<br><br>***Less likely:*** Early memory impairment, typical Parkinsonian resting tremor |

## Parkinson's disease dementia

PD is a well-known disease of motor function due to dopaminergic neuron loss in the substantia nigra. PD is also associated with mild cognitive impairment (MCI), and with a risk of dementia that is nearly 6 times that of age matched counterparts (Poewe et al, 2008). Estimates of the progression of MCI to dementia in PD are highly variable (from 5–100%) and seem to depend on heterogeneity in the study design. This degree of variation seems unhelpful, and further research is needed (Hu et al, 2014; Pigott et al, 2015).

Prevalence of cognitive impairment increases with illness duration, older age of onset of PD, movement disorder severity, poor cognitive baseline, and history of REM sleep behaviour disorder (RSBD). PDD is a risk factor for long-term care and increased carer burden.

### Neuropathology

PDD appears distinct from AD, both in cognitive profile and neuropathology. The pathological hallmark of PD, Lewy bodies, progresses in a predictable way, and the cortical spread may be linked to the severity of cognitive impairment (Braak et al, 2005). Cholinergic deficits appear to play a role in cognitive dysfunction.

### Clinical features

Early visuospatial and executive dysfunction are more common in PDD, in contrast to the prominent early short-term memory loss and language deficits seen in AD. Interestingly, impaired driving ability in mild to moderate PD has been associated with these early deficits, rather than with motor symptoms (Uc et al, 2007). When they do occur, memory impairments involve retrieval difficulties assisted with cues.

RSBD can occur years before the onset of PD or DLB. Visual hallucinations can occur in cognitively intact patients, but their presence (often of people/animals) increases the likelihood of developing a dementia (Fénelon et al, 2000). Personality changes, such as loss of emotional control, outbursts of anger and obsessive behaviours, may occur.

### Diagnosis

Distinguishing PDD from DLB is difficult; this is largely done on the basis of the temporal relationship of the motor and cognitive symptoms. Diagnosis requires the development of a dementia syndrome in the context of established PD. However, accurate diagnosis cannot be made without a detailed history. The cognitive decline in DLB may be more rapid, with earlier onset of delusions and hallucinations. The motor symptoms may be of greater severity in PDD, and tremor may be more common (Galasko, 2017).

## Dementia with Lewy bodies

DLB has characteristic Lewy bodies (abnormal deposits of alpha-synuclein in the cortex, brainstem and limbic regions of the brain) affecting neurotransmitter systems (particularly dopamine and acetylcholine), which give rise to clinical features.

DLB can overlap with AD, with a third to half of those with diagnosed AD revealing co-existing Lewy body pathology on post-mortem (Outeiro et al, 2019).

### Clinical features

In contrast to PDD, the dementia either precedes or occurs simultaneously with the motor symptoms. Its onset may be abrupt, with rapid progression. Falls may be an early presenting symptom. Memory can be considerably preserved compared with AD but, when present, demonstrates retrieval problems (Mormont et al, 2003). Patients can present with significant fluctuations in attention and

cognition (in contrast to PDD) and vivid visual hallucinations which may predate parkinsonism (Jellinger, 2018). Indeed, cognitive fluctuations and visual hallucinations form part of the increasingly-recognised syndrome of mild cognitive impairment with Lewy bodies (Donaghy et al, 2018).

Early impairments in attention, executive and visuospatial functioning often cause patients or others to notice a decline in work function or driving competency before more severe deficits appear. Parkinsonism can be as severe as PD, although it is usually milder, bilateral and symmetrical. People with DLB are often highly sensitive to antipsychotic medication. Hyposmia may be an early feature and can help to distinguish DLB from AD (Beach et al, 2020).

RSBD, including disturbed sleep wake cycle, nightmares, confusion and acting out dreams, may predate the diagnosis of DLB by several years. Daytime somnolence, long naps and episodes of staring and disorganised speech correlate better with DLB than AD (Ferman et al, 2004). Confusingly, patients may have autonomic dysfunction resembling that seen in MSA, although of a lesser severity (Thaisetthawatkul et al, 2004).

### Diagnosis

Probable DLB requires the presence of two symptoms of fluctuating cognition, visual hallucinations, extrapyramidal signs and RSBD. Specificity at autopsy is around 85%, which means that scoring positively on the criteria means the patient is fairly likely to have DLB (Outeiro et al, 2019).

## Management of PDD and DLB

There is no cure for PDD or DLB. Management of the Lewy body dementias (DLB and PDD) has been recently reviewed by Taylor et al (2020). There are now clinical trials and meta-analyses that provide evidence to guide treatment of cognitive, neuropsychiatric and motor symptoms. The cholinesterase inhibitors donepezil and rivastigmine are recommended for the treatment of the cognitive disorder of DLB (National Institute for Health and Care Excellence (NICE), 2018), and rivastigmine has been shown to be of some benefit in PDD (Rolinski et al, 2012).

The management of neuropsychiatric symptoms can be challenging. Psychotic symptoms can be attributable to the dementia or side effects of antiparkinsonian medication. Treating the debilitating movement disorder without worsening psychotic symptoms and cognition is a fine balance. Visual hallucinations are particularly distressing when they occur, and medication may be required. If antipsychotics are to be used, quetiapine or clozapine may be preferred, although the risks of vascular events and death remain. Medications, such as tricyclic antidepressants, benzodiazepines, anticholinergics and dopamine agonists, can impact on cognition, and patients should be managed on the minimum effective dose of PD medication. Melatonin or clonazepam may help to alleviate RSBD.

Non-pharmacological approaches (treating reversible causes, drug toxicity and environmental factors) are generally recommended as the first line of approach to behavioural and psychological symptoms, especially given the importance of these in other dementias and the lack of robust pharmacological interventions (Connors et al, 2018). Alternative approaches involving cognitive training and exercise may help improve cognition in this population (PDD), but further research is needed (Szeto and Lewis, 2016).

The often lengthy duration of these neurodegenerative conditions provide opportunities for nurses to have meaningful input into complex disease management, which not only means pharmacological management, but also having difficult conversations about realistic disease progression and the more practical aspects of planning for the future (Lennard, 2018).

## Parkinson-plus syndromes

Parkinson-plus syndromes are a group of neurodegenerative atypical Parkinsonian disorders.

### Multi-system atrophy

MSA affects the central and sympathetic nervous system and is both a synucleinopathy and tauopathy. MSA appears to be a spectrum of three disorders: cerebellar MSA (MSA-C), parkinsonian MSA (MSA-P) and olivopontocerebellar atrophy (OPCA).

The exact cause is unknown, but abnormal proteins may cause glial and myelin dysfunction and neurodegeneration (Jellinger, 2014). The degree of cognitive dysfunction may correlate with cortical involvement (Bak et al, 2005). It is rapidly progressive, and patients may be bedbound within 5-8 years (Tada et al, 2007).

### Clinical presentation

All subtypes have early autonomic symptoms; urogenital symptoms usually occur first. Others include orthostatic hypotension and erectile dysfunction. Urinary dysfunction also occurs in PD, but its occurrence in MSA is much earlier (within less than 2 years in one study) (Bloch et al, 2010).

Predictably, MSA-C has predominant cerebellar signs (ataxia, eye movement disorders, ataxic dysarthria), whereas MSA-P has predominant Parkinsonism. Olivopontocerebellar atrophy affects balance, coordination and speech.

RSBD is common, presenting in up to two-thirds of patients (Plazzi et al, 1997). Restless legs and daytime somnolence affects up to 30% of patients (Moreno-López et al, 2011). A reasonably specific feature for MSA is laryngeal stridor (Cortelli et al, 2019), initially occurring during sleep with daytime occurrence with disease progression. There is an association with sudden death (Silber and Levine, 2000). Hyposmia is less severe compared with PD (Kikuchi et al, 2011).

Cognitive function is often well preserved, but up to 20% of MSA patients may develop dementia (Brown et al, 2010).

Parkinsonian symptoms may occur before autonomic symptoms, which causes diagnostic difficulty, since MSA-P may appear identical to PD in the early stages. Autonomic failure can also occur in DLB and PD (Palermo et al, 2020). In DLB, the degree of cognitive impairment can be similar to that seen in MSA; therefore, DLB accompanied by autonomic impairment can be difficult to distinguish from MSA (Koga et al, 2015).

### Diagnosis

Probable diagnosis of MSA requires autonomic dysfunction (urinary incontinence or erectile dysfunction) or orthostatic hypotension and either parkinsonism poorly responsive to levodopa or cerebellar syndrome.

If possible, an accurate and early diagnosis is valuable, due to the rapid progression of the disease and the huge difference in life expectancy between PD and MSA. However, a definite diagnosis of MSA can only be made following post-mortem examination. The accuracy of clinical diagnosis varies between 60–78% (Koga et al, 2015; Miki et al, 2019). Neuroimaging is of some help, but a retrospective study comparing diagnosis in life with autopsy results showed 38% of MSA patients had normal neuroimaging (MRI) (Koga et al, 2015). The appearance of the pons on MRI is sometimes said to resemble a hot cross bun, but this sign is uncommon and nonspecific (Koga et al, 2015).

MSA patients can show an initial, but unsustained, response to levodopa (Parati et al, 1993), and they are more likely than PD patients to suffer side effects (particularly dyskinesias and orthostatic hypotension).

## Progressive supranuclear palsy

There are several variant phenotypes of PSP. The most common type is known as Richardson's syndrome. Other phenotypes consist of the same presentation combined with other clinical features, such as frontal or cerebellar signs.

PSP may be caused by a complex interplay of genetic and environmental factors (Litvan, 2004). The condition results in accumulation of tau proteins in neurofibrillary tangles, with degeneration of the nigral and subcortical areas.

### Clinical presentation

Patients tend to present with postural instability, early falls (usually backwards) and gait problems (stiff and broad-based). All variants display bradykinesia but, unlike PD, the slowing is without decrement.

PSP is associated with the development of a vertical supranuclear gaze palsy (people can move their eyes sideways but not up and down) and a pseudobulbar palsy. There may be a distinctive 'astonished' facial expression due to facial dystonia and oculomotor abnormalities (Litvan, 2004).

Cognitive impairment (often early and severe) is common and tends to involve frontal lobe dysfunction. Patients show difficulty processing information, executive dysfunction and reduced verbal fluency (Litvan, 2004). They may have speech and language impairments. Behavioural problems, such as anger, emotional lability and disinhibition, may be present.

Insomnia can be an issue, but RSBD is uncommon (Boeve et al, 2003). Olfaction is preserved. These traits and the cognitive impairment may help differentiate between PSP, PD and MSA.

The decline in functioning is usually rapid, resulting in a need for full care within 3-4 years.

### Diagnosis

As there are multiple variants of PSP, the clinical presentation can vary quite substantially, often leading to a late diagnosis. Diagnosis should be considered with rapidly progressive Parkinsonian signs, early falls, oculomotor deficits and poor levodopa response.

Diagnosis is clinical; neuroimaging can be supportive but is not diagnostic. It may show generalised and brainstem atrophy. An appearance known as the hummingbird sign may be seen on MRI, which is due to midbrain atrophy with preservation of the pons (Page and Gaillard, 2020).

There are standardised diagnostic criteria that identify possible, probable and definite PSP, and a definitive diagnosis relies on post-mortem identification of neurofibrillary tangles in the basal ganglia and brainstem (Hauw et al, 1994). 'Possible' PSP diagnosis requires either vertical supranuclear gaze palsy, or a combination of slowed vertical saccades and postural instability, causing falls in the first year. Probable diagnosis requires all three of these features.

## Corticobasal degeneration

In CBD, parts of the cerebral cortex and deeper brain regions, such as the basal ganglia, degenerate. Diagnosis antemortem remains low (25–56% of cases) (Armstrong et al, 2013). Prognosis is variable, but the median survival in two studies was 5.5 and 7.9 years (Wenning et al, 1998; Murray et al, 2007).

### Clinical presentation

Varying phenotypes cause varying symptomatology. Cortical and extrapyramidal symptoms may start unilaterally (usually affecting one limb) and spread bilaterally. Cognitive or behavioural changes may precede motor symptoms. Behaviours tend to reflect frontal involvement, with compulsive behaviour, sexual disinhibition, social withdrawal, apathy and unmotivated laughter being common.

Extrapyramidal signs include appendicular rigidity (which can be severe), less severe axial rigidity and limb dystonia (Wenning et al, 1998). Limb rigidity, bradykinesia or clumsy limb and postural instability are the most common presenting motor signs. The gait may resemble PD or be more wide-based. Tremor is less common in PSP than in PD, occurring in up to about 40% of cases, though it is often mild or intermittent, so may not even be observed (Fujioka et al, 2016).

Cognitive features include visuospatial impairment, apraxia (ideomotor and ideational), changes in speech (including dysarthria and aphasia). Episodic memory may be reasonably preserved, and memory difficulties are due to retrieval problems (Pillon et al, 1995).

With progression, there is eventual involvement of upper and lower extremities. The 'alien limb' phenomenon (patient feels their limb does not belong to them, has a mind of its own and is not under voluntary control) occurs in 30–50% of cases (Hanna and Doody, 2000). There may be some oculomotor dysfunction, with slow saccadic pursuit movements (Stover and Watts, 2001).

### Diagnosis

Neuroimaging may show asymmetric cortical atrophy and, in particular, fronto-parietal atrophy. The EEG may be abnormal with disease progression.

Definitive diagnosis is by postmortem examination showing asymmetric fronto-parietal atrophy, degeneration and depigmentation of the substantia nigra (Boeve et al, 1999). The cortex shows neuronal loss, achromatic swollen neurons and neurofibrillary tangles in a distribution similar to PSP.

However, making the diagnosis in life remains complicated, due to the variable clinical features of the disease, which do not always correlate with the underlying neuropathology (Lee et al, 2011). One study found that only 35% of those diagnosed in life were diagnosed with CBD postmortem (Lee et al, 2011).

A diagnosis of probable CBD requires an asymmetric presentation and at least two signs of limb rigidity or akinesia, limb dystonia, limb myoclonus and one of orobuccal or limb apraxia, cortical sensory deficit or alien limb (Armstrong et al, 2013).

### Management of Parkinson-plus syndromes

There is no curative or disease-modifying treatment for any of the syndromes, so management focuses on symptomatic treatment. They tend to respond poorly to dopaminergic drugs, although some cases may have temporary benefits, so a trial may be worthwhile. There may be troubling side effects, including visual hallucinations and dyskinesias.

In CBD, tremor and myoclonus may respond to benzodiazepines, and baclofen may help with rigidity. Dystonia and painful limbs may have some limited response to botulinum toxin.

The motor symptoms of MSA are difficult to treat, and there are no effective treatments for the cerebellar symptoms. Orthostatic hypotension can be treated with non-pharmacological measures, such as compression stockings or easing dietary salt restriction, or with medication, such as fludrocortisone.

The evidence base is limited, and many findings are extrapolated from Parkinson's disease studies; however, one recent systematic review highlights the importance of speech and language therapy (Tilley et al, 2016). Speech and language therapy, occupational therapy and physiotherapy can be useful in maintaining mobility for as long as possible. Swallowing difficulties may progress to such a stage that artificial feeding may be necessary.

The important contributions of nursing are considered below in the Discussion section of this paper.

## Huntington's disease

Huntington's disease (HD) is a genetic neurodegenerative disorder causing progressive movement, cognitive and neuropsychiatric symptoms, inherited in an autosomal dominant fashion; there is a 50% risk with one affected parent. The near perfect penetrance means that 'all carriers eventually become patients' (Alzheimer Europe, 2005). The mutation to the Huntington gene on chromosome 4 leads to an expansion of the CAG trinucleotide repeat. Successive generations often have an earlier age of onset, are associated with higher numbers of repeats, more rapid progression and a shorter life expectancy.

Life expectancy is around 20 years from onset. The symptoms and progression of HD remain devastating to patients and their families.

People living with HD may initially be able to live reasonably independently while experiencing relatively minor motor and cognitive deficits. With progression, this becomes less tenable as reliance on others due to cognitive and motor decline increases. The late stages may see patients bedbound.

### Clinical presentation

HD is diagnosed by the presence of chorea, a family history and a positive genetic test. Predictive genetic testing is available before the presentation of clinical symptoms, and the decision to undergo testing, either before or after symptom presentation, is a personal one fraught with moral and ethical dilemmas.

Motor symptoms change over the duration of the illness and are both hyper- and hypokinetic. Bradykinesia tends to occur early and increases throughout the duration of illness to akinetic rigidity syndrome. The well-known dance-like movement of chorea peaks around midway, and dystonia tends

to occur in the later stages (Burgunder et al, 2011). Eye movements, including the ability of the eyes to accurately jump between targets, may also be affected.

Inevitably, at some point patients are likely to experience neuropsychiatric symptoms (agitation, irritability, dysphoria, anxiety, apathy, psychosis, sleep disorders, impulsivity), either due to side effects of medication, the inherent neurodegeneration in the brain or psychological consequences of adjusting to and living with a diagnosis of HD. Suicide rates are 4-6 times that of the general population, and suicidal ideation is elevated in HD carriers (20%) (Wetzel et al, 2011).

Even early on, emotional recognition is impaired. Sufferers may have difficulties recognising their own actions and feelings, leading to family difficulties and challenging behaviours. They may exhibit impaired perception of time, spatial recognition and difficulty identifying smells. Often, they are unaware or in denial of their condition (anosognosia) (Wibawa et al, 2020).

Cognitive changes include executive dysfunction, problems with memory retrieval and difficulties with concentration and attention (Ghosh and Tabrizi, 2018). Decision-making and multi-tasking may be difficult. Patients may struggle with planning and sequencing, causing difficulties at work and home. Subtle neurological and cognitive signs (e.g. minimal choreiform movements, problems with memory and attention, or even changes in personality like becoming less considerate), as well as neuroimaging changes, may occur many years prior to clinical symptoms (Aylward et al, 2004).

## Management

Whilst HD dementia is incurable, there is some symptomatic treatment. Treatment can be difficult, due to the tendency of certain side effects to help one symptom and worsen another. That said, good management of symptoms and patient and carer education can delay institutionalisation and carer burnout. The importance of advance care planning cannot be emphasised enough. The ability to plan for the future, such as with an advance care plan or an advance decision to refuse invasive treatment, enhances an individual's autonomy, and nurses are well placed to assist with this process. With the establishment of a good therapeutic relationship, nurses are able to share the patient's journey, including ongoing discussion of hopes and wishes (Lennard, 2018).

The inherited nature of HD presents its own challenges. It often presents in middle age or younger, thus causing different problems to those with the condition, family and carers compared to, for example, the challenges of late onset AD. It is estimated that, for each person with the HD gene, a further 20 people are affected, due either to the significant dependence on caregivers or the genetic effects of inheritance (Domaradzki, 2015). Given the mode of inheritance, the implications of testing, a positive genetic test and diagnosis are significant and can be psychologically demanding. A person's

whole life trajectory can change in an instant: 'once the result of the test is disclosed, the individual and the family cannot live without the knowledge it brings' (Andersson et al, 2016).

### Neuropsychiatric symptoms

Cholinesterase inhibitors and memantine, which are used for AD, are ineffective in HD (Fernandez et al, 2000).

An understanding of the underlying causes of challenging behaviour (such as difficulties communicating needs), pain, environmental factors, underlying medical and psychiatric conditions is important, and *Table 2* shows some of the treatments used for frequently observed symptoms. Irritability is common and may be a combination of loss of impulse control, frustration with being unable to complete previously routine tasks, and unawareness. Expert consensus guidelines published in 2018 identify benzodiazepines, antidepressants and antipsychotic medications as potentially helpful (Anderson et al, 2018). Melatonin may aid sleep.

**Table 2: Medications often used in treatment of symptoms in Huntington's disease**

| | |
|---|---|
| Irritability | SSRIs; antipsychotics; mood stabilising anti-epileptics |
| Acute agitation | Benzodiazepines; antipsychotic medication |
| Persistent agitation | Antipsychotics; mood stabilising anti-epileptics |
| Anxiety | Antidepressants; antipsychotics |
| Apathy | Antidepressants; activation antidepressants; stimulants |
| Psychosis | Antipsychotics |
| Sleep disorders | Melatonin; hypnotics; sedating antidepressants; sedating antipsychotics |

## Discussion: the nursing role

Even in a brief account like this, it is apparent that these different dementia-causing disorders have their own distinct diagnostic, management and personal challenges. Unfortunately, apart from genetic testing in HD, there is no definitive diagnostic test; instead, accurate and timely diagnosis rests on a comprehensive clinical history and neurological examination. As several of these disorders are relatively uncommon and may have multiple phenotypes and symptoms that overlap with other

diseases, the diagnosis may not always be initially accurate. It may be that the correct diagnosis only emerges with time. Lewy body spectrum disorders and Parkinson-plus disorders may be initially misdiagnosed as PD, but unusual features, such as early falls, visual hallucinations, autonomic instability and oculomotor dysfunction, may indicate a rarer cause.

Nurses play an important role at different stages of dementia. This includes post-diagnostic support, signposting patients and their families to sources of information and advice, and helping people to review their changed situations (with regard to employment, driving, social activities, and managing their financial affairs, for example). It may also include offering anxiety management or other forms of psychological support, such as mindfulness, to alleviate distress among people with dementia and/or their families. Alongside lasting powers of attorney, advance care planning is important, as are respite, care packages and hospices in the later stages, to aid families and carers (Thompson et al, 2020). Nurses are often well-placed to act as care managers for the complex array of health and social care arrangements that may be needed for a person living with dementia and their family.

Particularly in relation to the Parkinsonian dementias, nurses may be well-placed to detect the emergence of new or unexpected symptoms, since they are more likely to spend more time, over a longer period, with patients and families. This may be helpful in flagging the possibility of DLB in a person with supposed AD, or in helping to detect early symptoms that suggest a Parkinson-plus condition. Doing so may help point to interventions of treatments that are helpful or which may reduce the risk of harm—from overuse of antipsychotic medication in DLB, for example.

Collectively, when it comes to these conditions, there are no curative or disease-modifying treatments, just symptomatic management. While timely diagnosis is not easy, these rarer causes of dementia should be considered. Specialist nurses are available for some. However, all nurses, (whether specialist Parkinson's disease nurses, Huntington's disease nurses, Admiral dementia nurses or generalist nurses) have a large part to play in providing appropriate, timely and joined-up care to both patients and those supporting them. With growing numbers of patients with dementia (particularly less common subtypes), generalist nurses may feel underqualified or out of their depth. Given the life-changing nature of a diagnosis of dementia and the positive impact that good nursing can have—from timely diagnosis NS support to patients and carers to the provision of appropriate information regarding future care planning to preventing avoidable emergency admissions—it is important that nurses are aware of how to acquire the necessary skills. Therefore, it is important that nursing education includes sufficient coverage of dementia and also that nurses have post-qualification access to educational and continuing professional development resources in relation to

dementia. The Higher Education for Dementia Network (2015) may be consulted for proposed curricula.

## Funding
No external funding was received for this work.

## Declaration of interests
The authors have no conflicts of interest to declare.

## Key points
Less common types of dementia cause 10-15% of all cases of dementia, and the most important are dementias with Parkinsonian features, Huntington's disease, frontotemporal dementia, human immunodeficiency virus (HIV), Creutzfeldt-Jakob disease and other prion diseases, and alcohol. This paper discusses Parkinsonian dementias and Huntington's disease.

Parkinsonian dementias include Parkinson's disease dementia (PDD), dementia with Lewy bodies (DLB), and Parkinson's-plus syndromes such as multi-system atrophy, progressive supranuclear palsy and corticobasal degeneration. PDD and DLB appear to be on a spectrum as to whether the earliest features are motor or cognitive in nature. DLB has a distinct pattern that can enable it to be separated from Alzheimer's disease, and it seems to have a more rapid course and worse outcomes.

There are no cures for these disorders, so the emphasis in on supporting patients and their families, and in dealing with symptoms and other problems as they arise. Nurses are often well placed to develop and manage care plans that adapt along with the progressive nature of these conditions.

## CPD questions
In your practice, have you seen patients with Parkinson's disease, especially those who have had it for several years? Think about what their mental state was like, for example, mood, concentration, memory. You may have noticed that their condition fluctuated and that treating their symptoms became more difficult over time.

Think about why it may be important to diagnose somebody as having dementia with Lewy bodies rather than having Alzheimer's disease. What might you need to explain to them and their family members about their symptoms and treatment?

Reflect on the problems that may arise from having rapid eye movement sleep behaviour disorder (RSBD). First think about what the patient may notice and then consider what it would be like to be trying to sleep in the same bed.

Unless you work in a very specialised clinic, you may not see many people with disorders like progressive supranuclear palsy or corticobasal degeneration. So how would you prepare yourself to provide care for them if you find yourself looking after a patient with one of these conditions? How much knowledge do you think you would need?

Consider the impact on a family if one of the adults is diagnosed with Huntington's disease. How many people are likely to be at risk of having the condition? What are the issues about taking a test for the presence of the HD gene?

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