Navigating Unchartered Waters: A Nursing Perspective on Lewy Body Dementia

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Abstract:

Although Lewy body dementia (LBD) is the second most common form of dementia to Alzheimer’s disease, this more rapidly progressive neurodegenerative disorder remains largely unknown to the public and is under-recognised by health care professionals.

Early and accurate diagnosis is complicated by many different presentations of the disease which can include a mixture of clinical features seen in both Alzheimer’s disease (AD) and Parkinson’s disease (PD). Motor and cognitive dysfunction as well as behavioural and mood disturbance are common overlapping features. What sets LBD apart however, is the variability and unpredictability of the disease and sensitivity to conventional anti-psychotic medications.

The ambiguous nature of LBD places significant stress on caregivers and presents unique challenges for the long-term clinical management of the disease. This paper is aimed at raising awareness of LBD and proposing key nursing interventions to enhance both quality and length of life.

Key words: Lewy Body Dementia, Parkinson’s Disease Dementia, Dementia with Lewy Bodies, Caregiver Burden, Nursing Care

Introduction:

Definition

“Lewy body dementia” (LBD) is broadly considered to consist of two related disorders—Parkinson’s disease dementia (PDD) and dementia with Lewy bodies (DLB) (Connors et al., 2017). Neuro-pathologically, these spectrum disorders are characterised by the widespread distribution of aggregated α-synuclein, forming structures called Lewy bodies in neuronal cell bodies and processes (Galasko, 2017; Stubendorf, Aarsland, Minthon & Londos, 2012). The abnormal clumping and accumulation of α-synuclein alters chemical processes causing a loss of functionality and eventually cell death (Fields, 2017; Perkins, 2017).

The diffuse distribution of pathology in LBD gives rise to a diverse and challenging range of symptoms including Parkinsonism, autonomic dysfunction, recurrent hallucinations, cognitive fluctuations, mood disorders, REM sleep behaviour disorder and moderate memory impairment (Jellinger, 2017; Killen et al., 2016; McKeith et al., 2017).

PDD vs. DLB Debate

Sharing of pathophysiological findings between PDD and DLB has sparked controversial debate regarding whether these are two distinct disease processes or whether they are the same entity that occurs on a continuum (Fields, 2017). A 1-year rule was proposed to split DLB and PDD; PDD is diagnosed if motor symptoms precede cognitive symptoms by more than a year; DLB is used if cognitive decline precedes or presents with the initial motor symptoms (Galasko, 2017).

Some consider the separation of the two entities as being useful to enhance diagnostic awareness and to encourage the study of both phenotypes so that one day their distinct pathophysiology can be better understood.
and lead to the development of disease-modifying treatments (Friedman, 2018; Galasko, 2017).

Others would argue that with disease progression, the course of the dementia, the symptoms and underlying brain changes are more similar than different. Therefore, with management being the same, regardless of diagnosis, clinically the most appropriate and helpful term to encompass these two overlapping syndromes is LBD.

**Early Diagnosis**

Early identification of patients at risk of severe cognitive impairment and dementia can help to inform choice of pharmacotherapy and assist with a personalised approach to treatment (Kehagia, Barker & Robbins, 2010). Distinguishing LBD from other dementias can be difficult particularly when shared Alzheimer’s (AD) pathology exists and often a misdiagnosis of AD is made (Galasko, 2017; Walker, Possin, Boeve & Aarsland, 2015).

A lack of awareness of typical symptoms, poor familiarity with the value of obtaining a diagnosis and variable general practitioner knowledge can cause delays in obtaining an accurate diagnosis which contribute to overall feelings of burden for caregivers (Jackson et al., 2016; Legett, Zarit, Taylor & Galvin, 2010). LBD is a more rapidly progressive disease than AD, early diagnosis allows caregivers and families to access interventions, plan for unexpected deteriorations in cognition, motor function and behaviour and utilise supportive resources (Galvin et al., 2010). Careful monitoring can assist with the prevention of a variety of complications and minimising exposure to medications that may provoke symptoms or cause potentially life-threatening conditions such as severe neuroleptic sensitivity (Mueller, Ballard, Corbett & Aarsland, 2017).

**Core Clinical Features:**

**Fluctuating Cognition**

Patients with LBD are prone to spontaneous alterations in cognition, attention, alertness and arousal including staring spells, confusion and incoherent speech (Galasko, 2017; McKeith et al., 2017). These episodes of fluctuating behavioural inconsistencies discriminate LBD from other dementias and are correlated with increased caregiver distress (McKeith et al., 2017; Mueller et al., 2017).

Planning activities in advance and scheduling rest periods may help to prevent tiredness and episodes of confusion (Londos, 2017). Large, randomized placebo-controlled trials show good evidence that the cholinesterase inhibitors (AchEI), Donepezil and Rivastigmine can improve cognition, agitation and attention (Stinton et al., 2015).

**Recurrent Hallucinations**

Cortical Lewy bodies in LBD are associated with complex recurrent visual hallucinations (Galasko, 2017). Vivid and well-formed false perceptions of people, insects or animals are difficult to manage because of their realistic details (Perkins, 2017; Walker et al., 2015). These phenomena are rarely reported on voluntarily due to variations in patient insight and emotional reaction and the suggestion that something might be wrong with them (Londos, 2017; McKeith et al., 2017). This can contribute to carer strain as a failure to disclose hallucinations delays treatment, affects quality of life (QOL) and can ultimately lead to hospital admissions.

The timely identification of hallucinations ensures access to pharmacological and psychosocial interventions (Tang, Burn, Taylor & Robinson, 2016). With careful blood monitoring for agranulocytosis, Clozapine may help to reduce hallucinations (Walker et al., 2015). Sertraline is well tolerated due to being short-acting but must be monitored for drowsiness (Galasko, 2017). Input from liaison neuropsychologists can assist with the design and implementation of treatment plans that can improve the management of this disturbing and destructive complication (Fields, 2017).

**Parkinsonism**

The probable or possible diagnosis of LBD requires one or more spontaneous cardinal features of parkinsonism: Bradykinesia (slowness of movement and in amplitude or speed), rigidity or resting tremor (McKeith et al., 2017). The management of motor symptoms in LBD is complicated by a poor response to dopaminergic treatments and an increased risk of psychosis (McKeith et al., 2017). Levedopa is commenced slowly and kept to the lowest effective dose with the least amount of adverse reactions (McKeith et al., 2017). Dopamine agonists can be problematic due to their high risk of provoking behavioural symptoms therefore carers and nursing staff must maintain careful vigilance. Patients at risk of falling should be referred to physiotherapy and occupation-
al therapists for safety assessments. Additionally, bone density screening and assessment of Vitamin D status should be done to minimise the risk of potentially life threatening fractures (McKeith et al., 2017).

**REM Sleep Behaviour Disorder**

REM Sleep Behaviour Disorder (RBD) is a parasomnia manifested by vivid dreams and complex motor movement (McKeith et al., 2017). Dreams often have a chasing, arguing or attacking theme where the patient is trying to protect themselves. RBD is thought to arise due to a lack of motor inhibition during REM sleep (Galasko, 2017). Unfortunately for the carer, sleep deficit occurs for them also and it may be necessary to change sleeping arrangements to avoid injury from flailing limbs. RBD can be managed with the use of Mirtazapine and can be complemented by low-dose Melatonin (Londos, 2017, Walker et al., 2015). In exceptional cases, Clonazepam may be useful with careful monitoring for hypotension. Patients and carers must be educated about improving safety to minimise injury (Walker et al., 2015) and the potential risks of sedation, falls, cognitive changes and agitation with prescribed treatments.

**Autonomic Dysfunction:**

Dysfunction of the autonomic system is common in LBD due to the a-synuclein pathology in central autonomic pathways (Galasko, 2017). Symptoms such as excess salivation and sweating can be distressing and embarrassing and erectile dysfunction inevitably impacts on intimacy.

**Hypotension & Syncope**

According to Barone et al., 2009 (as cited in Robertson et al., 2015) light-headedness, fatigue, generalised weakness, headache, nausea and impaired cognition can impact on independence and decrease QOL. Postural blood pressure should be checked routinely and side effects from anti-parkinsonian medications and antidepressants should be closely monitored (Stubendorff et al., 2012). Low-dose fludrocortisone may be prescribed after careful consideration and review of any concomitant anti-hypertensives. LBD patients tend to have a prolonged period of orthostasis after standing, therefore education about slow transitions from lying to sitting and standing position should be given. Additionally, where appropriate salt and fluid intake should be encouraged and compression stockings and abdominal binders may be considered (Walker et al., 2015).

**Urinary Incontinence**

Urinary incontinence contributes to sleep disturbance, social isolation and carer burnout. Management involves regular toileting to prevent anxiety and incontinence and forward planning for travelling. Referral to a continence nurse for continence aids and advice can be helpful. Oxybutynin and Solifenacin succinate may be prescribed to prevent urgency, frequency and leakage however, the potential benefit must be weighed against the risk of worsening cognition and delirium (Walker et al., 2015).

**Constipation**

Constipation in LBD is thought to be linked with Lewy body cholinergic impairment of the enteric nervous system (Lepkowsky, 2017). A weakening of the bowel muscles, poor muscle coordination and anal rectal changes make it difficult for bowel motions to occur (Vanderbergriendt, 2017). Encouraging a balanced diet, exercise and fluids is essential for prevention. Patients can be guided by their clinicians about the best use of laxatives, stool softeners and probiotics.

**Cognitive Decline:**

**Executive Dysfunction**

Deficits with working memory, set shifting and planning make it difficult to multi-task, and follow conversations and directions. Distraction and the loss of train of thought during household sequential tasks can have implications for personal care and safety within the home and ultimately leads to an early loss of independence.

**Attention**

Fluctuations in attention and mental flexibility are associated with driver difficulty (Fields, 2017). Slow reaction to sudden changes and a decreased ability to identify landmarks and traffic signs can contribute to at fault safety errors (Uc, Rizzo, Anderson, Spark. Rodnizky & Dawson, 2006)

**Visual Spatial Disability**

Depth perception and judgement of distance are altered in LBD. Severe difficulties with visuospatial functioning can predict rapid decline and the development of hallucinations (Walker et al., 2015). Clumsiness, trips, falls, mishaps around the home and misjudging distances are sensitive issues to report but need to be explored to inform treatment choices and ensure patient safety.
Mood and Personality Changes:

Anxiety

Fluctuating cognitive and motor function in LBD can contribute to worry, phobias and panic attacks. Heightened levels of anxiety can worsen parkinsonism leading to the avoidance of activities, social and emotional isolation and poor QOL (Fields, 2017). New situations and surroundings can cause confusion and panic which places limits on socialising and holidaying. These unpredictable mood changes can be a major stressor for carers and family members. The provision of education and early referrals for respite support can help to alleviate carer burden.

Benzodiazepines, such as Diazepam and Lorazepam can be prescribed cautiously for their calming effect with careful monitoring for sedation, confusion and paradoxical agitation.

Apathy

Reduced initiative and motivation combined with overwhelming fatigue and hypersomnia can be some of the most disabling features of LBD. Serotonin norepinephrine reuptake inhibitor (SSRI) anti-depressants may be worth a try and can be combined with Modafanil as a stimulant (Bomasang –Layno, Fadland, Murray & Himelhoch, 2015).

Depression

The unpredictability of LBD makes it a very isolating condition with patients often feeling like they are trapped in their own bodies. During times when insight and clarity are retained, patients can become frustrated with the increasing perception of burden. Impulsivity, disinhibition and emotional lability can contribute to awkward interactions and contributes to high dependency on spouses and carers and restrictions in social and living arrangements (Mueller et al., 2017). Venlafaxine and Melatonin can be used to improve sleep and reduce depressive symptoms (Londos, 2017). SSRIs such as Sertraline and Citalopram are most commonly used due to safer tolerability (Perkins, 2017) however, Fluoxetine and Paroxetine may also have a role in managing anxiety and depression with careful monitoring for gastrointestinal side effects.

Neuropsychiatric Complications:

Illusions

Objects, shadows and patterns can often be misinterpreted as people or animals (Galasko, 2017) and these misperceptions can have implications for mood and safety depending on a patient’s reaction to these distorted visual stimuli.

Delusions

There is often a paranoid theme to delusions in LBD with a focus on spousal infidelity, theft and home intruders (Goldman et al., 2014; Perkins, 2017). Capgras Syndrome is the most common unusual delusional syndrome where the spouse or close relative of a patient is believed to have been replaced by an imposter (Perkins, 2017; Moro et al., 2013). Reduplicative Paramnesia occurs when it is believed that a place simultaneously exists in 2 or more physical locations (Devinsky, 2009).

The reoccurrence of these symptoms contributes to carer burden which ultimately leads to more frequent hospital admissions, longer length of stay and accelerated nursing home placement compared to AD (Mueller et al., 2018). Clinicians need to be aware of these rare conditions so that factors such as pain, infection and interpersonal and environmental triggers that impact on delusions and agitation can be treated appropriately (Galasko, 2017; Moro et al., 2013). Carers should be encouraged to utilise family and community supports for regular scheduled respite to prevent carer burnout. A reduction of higher doses of levodopa may lead to an improvement in symptoms (Galasko, 2017; Moro et al., 2013). Case reports have shown that Gabapentin can reduce the symptoms of agitation (Stinton, et al, 2015). Pimavanserin recently received FDA approval for hallucinations and delusions associated with PD psychosis (Cummings, Isaacson , Mills, Williams, Chi-Burris, Corbet, Dhall & Ballard, 2014).

Care Planning:

In LBD cognitive decline, greater caregiver burden and a greater effect on quality of life seems to be more accelerated than in AD. These circumstances need to be considered when contemplating long-term treatment (Mueller et al., 2017). The extent of the burden on people with LBD and their carers calls for the development of comprehensive and individualised care plans including pharmacological and non-pharmacological
interventions (Morrin et al., 2018). As the disease progresses, this complex, multi-system disease requires multiple providers and ongoing communication and collaboration.

Nurses as often the first point of contact, are in a unique position to assess the need for caregiver support and education and to refer to social networks, community organisations and aged care and disability assessment teams for personal care, respite and permanent care approvals.

Early referral to allied health professionals for safety concerns can assist with keeping patients in the home for as long as possible. Social workers can assist with understanding government carer entitlements and the legalities of Patient Consent, Living Wills, Power of Attorney and Guardianship. Furthermore, nurses can help carers and families prepare for emergencies by providing education about the disease and medication complications and empowering them to be able to advocate for the patient in acute care settings where LBD may be poorly understood.

To alleviate any pressure on family members to make difficult decisions during a crisis, early discussions about Advanced Care Directives and long-term care preferences should be conducted and documented while the patient has insight and can have input into their future care.

Since patients may not always be forthcoming about their level of functioning and carers may be reluctant to speak out of turn, clinicians need to be astute in their assessment of their patient’s situation. Questions regarding personal care, managing medication and finances, driving, falls, safety in the home, exercise, cognitive stimulation and social interaction should be included in consultation to gain further insights into the day-day issues.

Conclusion:

In the absence of any cure or protective therapies for LBD, we need to take a wider approach to research and the provision of specialty health services.

The poor prognosis and high burden of care call for new treatment studies for LBD to be prioritised (Mueller et al., 2017). A better understanding of the pathophysiological mechanisms and prodromal states of these diseases may lead to more accurate diagnosis and the development of disease modifying and symptomatic treatments. Eligibility criteria need to be considered so that trials can be designed specifically to investigate LBD and expand the evidence base (Morrin et al., 2018). The development of comprehensive clinical guidelines for the management of LBD will require robust interventional trials evaluating the efficacy of combined pharmacological and non-pharmacological interventions to inform best practice (Morrin et al., 2018).

By raising awareness in the general community and developing more support resources specific to LBD, this may help to generate interest and understanding and reduce the stigma associated with this more rapidly progressive but lesser known form of dementia (Legett et al., 2010). Additionally, addressing the gap in knowledge of clinicians, patients and caregivers should be prioritised to ensure patients receive timely and appropriate treatments and support to be able to face the unique challenges of navigating these largely unchartered waters.

Disclosures:

Madelaine Ranola has served as an advisor for Pfizer and Global Kinetics and has received honoraria from Merck Sharpe and Dohme outside the submitted work

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Vale: Sr. Elizabeth Naigaga

Last month the Neuroscience Nursing community lost a great and inspiring nurse. Elizabeth Naigaga was the head nurse of the first ICU at Mulago National Referral Hospital in Kampala, Uganda. Through her tenacity and perseverance she strived to improve both care and facilities in Uganda, including the development of a neurosurgical high dependency unit. It was through her mentoring and leadership that she continued to improve care and services at the Mulago ICU.

In 2013, some of us were lucky enough to meet and hear Elizabeth present at the 11th Quadrennial WFNN Congress in Gifu, Japan.

Elizabeth lost her battle with cancer in May. This, the loss of a truly incredible, dedicated and passionate neuroscience nurse.

Elizabeth's passion to change the world she lived in will be honoured by a creating a scholarship fund in her name to benefit an African nurse with limited resources to attend an international nursing conference with WFNN, NCS or AANN. (Original story https://wfnn.org/)