Meeting Highlights

Diagnosis and Management of Dementia with Lewy Bodies – Challenges and Current Practices

Highlights of a GE Healthcare-sponsored educational meeting involving healthcare experts from across Europe, held in London, UK on the 23 March 2018

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Dementia with Lewy bodies (DLB) is regarded as the second most common cause of neurodegenerative dementia in older people. Despite this, DLB remains a challenging condition to diagnose, largely due to a varied presentation of symptoms that overlap with other conditions such as Alzheimer’s disease and Parkinson’s disease dementia. Achieving an early and accurate diagnosis of DLB is an important clinical goal, as it can have substantial benefits to both patient and caregiver. These include improved clinical outcomes such as cognition, patient quality of life, time to nursing home placement and mortality, as well as reducing caregiver burden and helping them to apply effective management practices and seek appropriate support for the patient. However, while current diagnostic criteria for DLB have been widely accepted, they are not always adhered to in clinical practice and/or fully understood. Strategies are required to increase awareness across the healthcare spectrum not only of DLB as a potential diagnosis, but also of its key features, diagnostic criteria, and the vital role of imaging techniques.

Keywords
Dementia with Lewy bodies, diagnostic criteria

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Here we report the key insights from a GE Healthcare-sponsored educational meeting on DLB, involving European healthcare experts, which was held in London, UK on 23 March 2018. Delegates in attendance represented countries from across Europe, including France, Germany, Italy, the Netherlands, Nordic countries, Spain and the UK. The main objectives of the meeting were: (i) to share and discuss current experience in the diagnosis of DLB and the management of patients with DLB across Europe; (ii) to discuss and learn best practice in the diagnosis of DLB; and (iii) to highlight the implications of an early and accurate diagnosis of DLB on patient management and outcomes.
As recently as the 1980s, dementia was typically defined as either Alzheimer’s disease or vascular dementia. However, with the development of more sensitive immunohistochemistry techniques it was subsequently discovered that a considerable proportion (up to 25%) of patients with dementia presented with some degree of Lewy body pathology at autopsy.2,11 As a result, the DLB Consortium was formed in the 1990s to generate diagnostic criteria for DLB. The first DLB Consortium consensus guidelines for the clinical and pathological diagnosis of DLB were first published in 1996.7

As new technologies, research and clinical insights applicable to DLB have emerged over the years, the DLB Consortium has reviewed and revised the diagnostic criteria, firstly in 2005 and most recently in 2017.2,11 While the 2017 criteria remain largely consistent with previous versions, key differences include: (i) a better differentiation between clinical features and biomarkers, with rapid eye movement (REM) sleep behaviour disorder (RBD) being upgraded to a core clinical feature and the list of supportive clinical features being extended (Table 1); (ii) dopamine transporter single photon emission-computed tomography (DaT-SPECT), metaiodobenzylguanidine (MIBG) cardiac scintigraphy, and polysomnographic confirmation of REM sleep without atonia now classified as indicative biomarkers (Table 1); and (iii) increased clarity about details of the patient examination and interviews.2 The latter includes questions relating to REM sleep (exclusion of confusional awakenings, severe obstructive sleep apnoea and periodic limb movements), visual hallucinations (well-formed, usually featuring people, children or animals) and cognitive and attentional fluctuations (daytime drowsiness, lethargy, staring into space or episodes of disorganised speech).2

**Diagnostic criteria**

“Updates to the DLB diagnostic criteria are a process of evolution, rather than revolution” – Ian McKeith

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**Diagnostic challenges and variations in clinical practice**

“One of the main problems with diagnosing DLB has to do with knowledge; even amongst neurologists or geriatricians very few know the exact criteria for a diagnosis” – Alessandro Padovani

Overall, despite the availability of defined diagnostic criteria, there remain challenges to achieving a timely and accurate diagnosis of DLB. These broadly fall into three categories: the nature of the condition (heterogeneity and a varied presentation), limited knowledge (of the condition or diagnostic criteria), and different approaches to clinical care between centres and countries. While advances in technology, clinical research, and efforts to increase awareness of DLB may help address the first two challenges, it is also important to identify the key differences in clinical care pathways between countries, to determine the best possible practice and thereby optimise patient outcomes. This is especially important for European countries where no national dementia plan currently exists.

Below is a summary of the distinguishing aspects of DLB diagnosis and clinical management, including clinical/biomarker assessments, specialist involvement and/or cost considerations, as outlined by the expert faculty for their respective countries during the DLB educational meeting on the 23 March, 2018.

**Germany**

In Germany, there is currently no national dementia plan and a diagnosis of DLB is mainly made by office-based neurologists or psychiatrists in a memory clinic or specialised movement disorder clinic, rather than by general practitioners. Indeed, most patients referred to memory clinics will have received little neuropsychometric testing. Notably, there are considerably more diagnoses of DLB in geriatric clinics compared with memory clinics, suggesting differences in either the visiting patients or in the diagnostic approach between the treatment centres. Unlike most other European countries, in the faculty’s experience, fluorodeoxyglucose positron emission tomography (FDG-PET) is often the preferred functional imaging tool used in a differential diagnosis of DLB, despite it being more expensive than magnetic resonance imaging (MRI) and cerebrospinal fluid (CSF) biomarkers, and of a similar cost to DaT-SPECT. Overall, the faculty noted that including DaT-SPECT (as well as FDG-PET) into the standard diagnostic work-up may be an expensive approach, potentially adding substantial incremental costs per quality-adjusted life year gained in patients with DLB.

**Italy**

In Italy, a national dementia plan has been in place since 2014, which was developed by the Ministry of Health in co-operation with different regions, the National Institute of Health and national patient/carer associations.12 There is also an organisation of specialised dementia centres (the Centres for Cognitive Decline and Dementia) and an Italian DLB study group to drive forward clinical research in DLB. In a recent questionnaire survey by the Italian DLB study group, it was shown that the vast majority of Italian dementia centres (91% of 135 centres) considered clinical and neuropsychological assessments to be the most relevant procedures for a diagnosis of DLB.10 The faculty recommended that prior to referral to a specialist centre, patients presenting with dementia should receive blood testing, an electrocardiogram (ECG) and computerised tomography (CT) scans. When a patient does present at a dementia centre with behavioural, visuospatial, cognitive or parkinsonian symptoms, MRI is usually performed, as well as psychological, neurological and psychiatric evaluations. If these are not conclusive, DaT-SPECT or FDG-PET should then be performed, dependent on the clinical picture. In line with this, the survey also showed that most Italian dementia centres have access to MRI (95%) and electroencephalography (EEG; 93%) facilities, but fewer centres have access to SPECT (75%).11

**The Netherlands**

In the Netherlands, patients diagnosed with dementia will usually be referred to a memory clinic where a standardised work-up is performed, usually consisting of a clinical examination by either a geriatrician, neurologist or psychiatrist; and cognitive testing and brain imaging (typically MRI). There are, however, four specialised Alzheimer centres where more elaborate diagnostic procedures can be performed. Patients visiting the specialised academic memory clinic at the VU University Alzheimer Centre in Amsterdam, for example, receive a full ‘day assessment’ involving a multidisciplinary approach (neurologist, psychiatrist, geriatrician and specialised nurse) and receive a variety of assessments including MRI, EEG, neuropsychological test battery, blood tests and a lumbar puncture. A clinical diagnosis is then reached by consensus in a multidisciplinary meeting.

In addition to the standard MRI and EEG assessments, which are inexpensive to perform and widely available in specialised centres, CSF analysis can provide useful information for a diagnosis (but not diagnosis) and DaT-SPECT, MIBG, polysomnography and FDG-PET may also be available to aid diagnosis in more complex cases. However, including additional biomarker analyses such as DaT-SPECT and FDG-PET into the standard diagnostic framework would substantially increase the associated costs.
Pharmacological management of DLB in the Netherlands is generally in line with the recent DLB Consortium recommendations, focusing on the cognitive, psychiatric, motor and other non-motor symptoms that form the core features of the disorder. Cholinesterase inhibitors are used to improve cognition and global function, rivastigmine/lower-dose clonazepam for hallucinations (haloperidol is excluded as it can worsen the condition), levodopa for Parkinsonian symptoms, clonazepam/melatonin for RBD, and selective serotonin reuptake inhibitors for depression/anxiety.

Spain
In Spain, there is no current national dementia plan; while the Spanish Neurological Society does grant accreditation status to dementia clinics, there is no actual recognition from the Spanish Health authorities. As such, dementia clinics are not well established across all 17 autonomous regions. In fact, they act as a ‘third step’ in the evaluation of patients with cognitive deficits: patients are typically referred to the clinics from general neurology centres. In contrast, movement disorder clinics are well established across Spain, although the majority do not have dedicated cognitive–behavioural assessment capabilities.

To aid the diagnosis of DLB, SPECT and EEG are widely available, but access to PET imaging, CSF analysis and MIBG assessments are limited and polysomnography may take up to 1 year. MRI is also widely available, though there is usually no specific image acquisition protocol for neurodegenerative disorders. A further challenge to diagnosing DLB in Spain, is reimbursement. A diagnosis of Alzheimer’s disease or Parkinson’s disease dementia is required for reimbursement of most therapies and activations of special care. In many cases this may lead to a diagnosis of these conditions in order to obtain proper care or reimbursement for patients presenting with dementia.

UK
In the UK, most National Health Service (NHS) Trusts have an agreed dementia care pathway; however, there are three different models operating depending on locality. In the first model, patients are assessed...
by “care facilitators” – primary care workers trained to perform assessments of people with cognitive decline who are not trained medical/nursing staff. This is efficient and inexpensive, but as part of the assessment no physical examination is performed, only 50% of patients receive a dementia blood screen, 25% receive an ECG and only 50% receive any form of brain imaging. In the second model, registered nurses work alongside a consultant within primary care. This is more expensive than the first model but involves a more comprehensive assessment that includes a physical examination, brain imaging in the majority of cases (CT, MRI), and additional investigations if indicated (DaT-SPECT, FDG-PET).

The third model involves patient assessment in a specialised memory clinic (performed by a doctor, registered nurse or clinical psychologists), is associated with midway costs, and includes a comprehensive medical history, a physical examination in most cases, brain imaging (CT/MRI), and additional investigations if indicated (DaT-SPECT, FDG-PET). The second model involves patient assessment in a specialised memory clinic (performed by a doctor, registered nurse or clinical psychologists), is associated with midway costs, and includes a comprehensive medical history, a physical examination in most cases, brain imaging (CT/MRI), and additional investigations if indicated (DaT-SPECT, FDG-PET). Additional biomarkers such as DaT-SPECT and FDG-PET are available for more complex cases.

### Early and accurate diagnosis – benefits to the patient and caregiver

“Giving an early and appropriate diagnosis helps to relieve the stress of the caregiver and anticipates the care that the patient will need”

– Guillermo Garcia-Ribas

“With a DLB diagnosis you can avoid multiple visits to A&E, avoid multiple visits to a GP, and start appropriate treatment so that the patient’s symptoms may improve”

– Zuzana Walker

“This is attended by the patient and caregiver. It has been noted that patients with DLB generally have a worse prognosis than patients with Alzheimer’s disease.14–16 As such, distinguishing DLB from Alzheimer’s disease and other dementias is an important treatment goal. Not only does it enable the timely use of appropriate pharmacological (and non-pharmacological) therapies,1 thereby improving cognitive, behavioural and delirium outcomes, it also prevents the use of inappropriate and harmful therapies such as antipsychotics (e.g. haloperidol)17 that may lead to increased hospitalisations and early nursing home admission (Case 1).15–16

An early diagnosis of DLB can also reduce hospital admissions (usually due to falls/fractures; Case 1) and shorten the duration of any hospital stays by enabling the effective management of the condition and more targeted care focussed on specific hospitalisation triggers.18 It may also facilitate the pro-active management of complications,10 and reduce caregiver burden, by helping caregivers appreciate the condition and treatment regimen, understand and better cope with patient’s symptoms, and mobilise appropriate healthcare services (e.g. a care co-ordinator, social care). Lastly, the ability to more accurately diagnose patients with DLB would improve the selection of patients for clinical trials, enabling further research and development of novel treatments to improve patients’ outcomes.

### Current best practice and future diagnostic improvements

Below is a summary of the key points from group and expert discussion sessions during the DLB educational meeting on the 23 March, 2018, focussed around current best practice for the diagnosis of DLB and potential ways to improve it. Discussions included representatives from across Europe, including France, Germany, Italy, The Netherlands, Nordic countries, Spain and the UK.

#### Case 1 – presented by Zuzana Walker

- **A 79-year-old patient with a complex presentation visited a UK memory clinic, where an initial diagnosis of vascular dementia was made.**
  - The daughter had noted cognitive changes over 1 year, with the patient forgetting entire conversations, having word-finding difficulties and not recognising friends.
  - The patient was independent in activities of daily living, had no delusions, no visual hallucinations, but had reported hearing voices they suspected were their grandchildren.
  - MRI revealed age-related generalised atrophy with relatively preserved medial temporal lobes and evidence of small vessel ischaemia.
  - The patient developed persistent delusions over the next 18 months, and was started on neuroleptic therapy with quetiapine 25 mg. Further rapid deterioration of the patient resulted in early admission to a care home and repeated hospital admissions due to falls.
  - During a hospital admission, symptoms of parkinsonism were noted by the liaising psychiatrist (bradykinesia and rigidity) and the patient was diagnosed with DLB.
  - Once the DLB diagnosis was made, the patient was housed in a care home, neuroleptic therapy was stopped, cholinesterase inhibitor treatment initiated and the patient showed improvement and stabilisation.
  - This highlights the importance of an accurate diagnosis of DLB, particularly with regard to the appropriate use of neuroleptic and cholinesterase inhibitor therapy, on patient outcomes.

#### Case 2 – presented by Alessandro Padovani

- **A 68-year-old patient presenting at an Italian neurology centre was given an initial diagnosis of mild cognitive impairment probably due to Alzheimer’s disease.**
  - The patient reported difficulties in concentration for 6 months, and the spouse reported that sometimes the patient was “not there”, forgot the date, did not recognise relatives or became confused.
  - There was no impairment in motor symptoms; non-motor symptoms were not assessed.
  - CT scan showed no hippocampal atrophy.
  - Cognitive testing showed amnestic multi-domain mild cognitive impairment with minor impairment in activities of daily living.
  - No treatment was initiated due to minimal impairment in activities of daily living.
  - Over the following 4 years the patient developed not only cognitive deficits and activities of daily living impairments, but also parkinsonian symptoms and hallucinations.
  - Following further clinical examination, MRI and DaT-SPECT, a diagnosis of DLB was made.
  - Subsequent medical history revealed that RBD, urinary dysfunction and hyposmia were present prior to the original diagnosis.
  - This highlights the need to consider DLB and take appropriate medical history during a differential diagnosis of dementia, something which may have enabled pro-active treatment of parkinsonian and psychiatric symptoms in this case.
Case 3 – presented by Guillermo Garcia-Ribas

- A 66-year-old patient was referred to a specialised centre with memory complaints. The clinical presentation and neuropsychological examination suggested a diagnosis of Alzheimer’s disease.
  - The patient reported memory problems for 3 years, including difficulty with word-finding and remembering names, transient disorientation, clumsiness and sleep problems.
  - The patient exhibited no rest tremor, reported no visual hallucinations, had a slightly impaired memory test score (41/50) and a severely impaired clock-drawing test (1/7).
- However, a subsequent letter from the patient’s spouse revealed the presence of sleep disorder, behavioural problems, and more severe memory and spatial orientation problems (forgetting where they are and what they were saying).
- This altered the direction of the investigations, with DaT-SPECT, polysomnography and MRI excluding Alzheimer’s disease and leading to a diagnosis of DLB.
  - This highlights the need for clear and directed interviews during diagnosis, as many core features of DLB are not self-reported.

Best practice

"Adhere to the criteria" – Richard Dodel

"DLB is a complex issue and a patient with DLB needs to be seen by a specialist familiar with DLB", "Whenever possible, additional biomarker evidence should always be obtained" – Ian McKeith

Differential diagnosis

When a patient presents with possible dementia, DLB should always be considered in the differential diagnosis. As DLB is a complex condition, adherence to the current diagnostic criteria is essential and expert involvement in both the diagnosis and clinical management should occur as early as possible. Ideally, a multidisciplinary approach (from nurses to specialists) should be employed, together with the use of screening interviews/questionnaires, as many of the core features may not be self-reported (Case 2 and Case 3).

Assessments

Suspected cases of DLB should receive a specialised clinical assessment, including a full neurological examination. DaT-SPECT has been shown to be more specific than clinical diagnostic criteria for DLB and should be the modality of choice to differentiate DLB from Alzheimer’s disease. However, in up to 10% of cases DaT-SPECT results may be normal even though the clinical picture is typical of DLB. In such cases, Lewy body pathology is probably more dominant in the cortex than the brainstem, and other imaging techniques such as MIBG may be required to confirm a diagnosis of DLB. Medial temporal lobe atrophy can be visualised using structural MRI and a lower rate of atrophy is typically seen in DLB compared with Alzheimer’s disease. However, this feature is of limited value in discriminating the two disorders and is therefore included as a supportive biomarker in the current diagnostic criteria. If medial temporal lobe atrophy is observed in a patient with DLB it probably indicates significant Alzheimer pathology and a worse prognosis. The same holds for CSF biomarkers. To date there are no DLB-specific biomarkers, but positive Alzheimer’s CSF biomarkers observed in patients with DLB may be associated with more rapid cognitive decline and a worse survival rate. Data from clinical studies suggests that FDG-PET has a lower sensitivity (70%) and specificity (74%) than that required for an indicative biomarker, therefore it is currently considered a supportive biomarker.

EEG biomarkers are also considered supportive because current evidence is only from studies with small sample sizes.

Clinical management and follow-up

Because of the potential risk of a severe sensitivity reaction, antipsychotic treatments should be avoided where possible. Current DLB Consortium guidelines propose the use of low-dose quetiapine, clozapine or pimavanserin as potential low-risk options for antipsychotic therapy, but clinical efficacy in DLB for all these agents has yet to be established.

DLB has a worse prognosis than Alzheimer’s disease, with faster cognitive decline, increased mortality, earlier nursing home admission and frequent and longer hospitalisations (resulting in greater financial burden). In addition, the spectrum of neuropsychiatric and behavioural symptoms common in DLB result in a lower patient quality of life and place a substantially higher burden on the caregiver compared with Alzheimer’s disease. As such, regular specialist follow-up visits should be performed to assess the effectiveness of patient care and treatment and adjust if necessary. Ideally, a potential post-diagnosis visit schedule should be 3, 6, 12 months and every year thereafter.

Ways to improve diagnosis

Improving the performance and reporting of biomarkers identified by the DLB Consortium as being clinically useful for a diagnosis of DLB (e.g. DaT-SPECT, polysomnography and MIBG) through training and education is a clear way to optimise their use in clinical practice and improve the diagnosis of DLB. Potential strategies to achieve this include the development of a formal algorithm and/or workflow detailing the sensitivities and specificities of different assessments (to enable selection of the most appropriate biomarker for any given situation), and nuclear medicine educational initiatives and/or quality assessments specific to DLB.

The development of novel cognitive tests and the refinement of diagnostic criteria to distinguish between DLB, Alzheimer’s disease and other dementias is an evolving process, and the more widespread use of these tests/criteria may improve the sensitivity of future diagnosis. Current examples include the pareidolia test, which uses DLB patients’ visuo-perceptual deficits and generates illusory phenomena similar to visual hallucinations, and ongoing work to refine the definitions of fluctuation and improve the characterisation of psychiatric symptoms in patients with DLB beyond visual hallucinations alone.

Perhaps the largest barrier to an early and accurate diagnosis of DLB is awareness, both of DLB as a potential diagnosis and of the current diagnostic criteria. This applies not only in primary care but also across the entire healthcare spectrum (e.g. primary care, psychiatrists, geriatricians, general neurologists, patient associations) – even in specialised memory clinics DLB is not always foremost in the mind when considering the differential diagnosis of a patient with dementia. Potential initiatives to achieve this include: (i) guidance/education on the core clinical symptoms (e.g. RBD, visual hallucinations, cognitive or attention fluctuations and motor parkinsonism, which may often be very mild) for physicians, potentially via diagnosis cards; (ii) further clinical research and publications; (iii) outreach to primary care and the public, e.g. via activities such as the ‘National DLB day’ observed in the Netherlands or by the formation of a European DLB Council; (iv) lobbying of politicians to increase awareness that this is a rapidly progressive condition and that effective management could reduce healthcare burden and improve patients’ quality of life; and (v) to include DLB as a potential component of dementia awareness programmes that are already in operation in many countries.
Conclusion

An early and accurate diagnosis of DLB can have substantial benefits to the patient and caregiver. These include: (i) being able to apply effective and appropriate treatments to improve patient outcomes such as cognition, quality of life, time to nursing home placement and mortality; and (ii) reducing caregiver burden, as well as helping caregivers to understand and manage the condition and seek appropriate support for the patient.

Further education in DLB is still required across the healthcare spectrum, to increase awareness not only of DLB as a potential diagnosis, but also of the key features, diagnostic criteria, and key role of biomarker imaging. Imaging techniques have shown good utility in the diagnosis of DLB over the past 10 years, with DaT-SPECT, MIBG cardiac scintigraphy, and polysomnography all now classified as indicative biomarkers.

Looking forwards, the ability to detect prodromal DLB may further enhance patient care and the management of DLB, and diagnostic criteria for prodromal DLB are currently in development by the DLB Consortium.