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EDITORIAL

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Revision of assessment toolkits for improving the diagnosis of Lewy body dementia: The DIAMOND Lewy study

As part of the UK National Institute for Health Research DIAMOND Lewy Programme (improving the DIagnosis And Management Of Neurodegenerative Dementia of Lewy body type), we have reported in this journal the development of two assessment toolkits to assist in the recognition and diagnosis of Lewy body dementia.[1] The "Assessment Toolkit for Dementia with Lewy Bodies" is for use by clinicians in memory and dementia services; the "Assessment Toolkit for Lewy Body Dementia," which facilitates an accurate diagnosis of either Parkinson's disease (PD) dementia or dementia with Lewy bodies, is designed for clinicians in movement disorder and geriatric medicine services.

The toolkits were developed to be easy to use by clinicians and to align with consensus diagnostic criteria for these dementias. Since our report appeared, the Fourth Consensus Report of the DLB consortium on the diagnosis and management of DLB has been published.[2] We have therefore updated our toolkits to align them with the new criteria and here summarise these changes. The link below takes you to our original paper, where the development of these toolkits is described (http://onlinelibrary.wiley.com/doi/10.1002/gps.4609/full) and which is free to download. The revised toolkits are in the Appendices to this Editorial.

1 | CHANGES IN DLB DIAGNOSTIC CRITERIA

Diagnosis of DLB according to previous 2005 criteria relied on the identification of core features of DLB (fluctuating cognition, recurrent complex visual hallucinations, and one or more spontaneous cardinal features of parkinsonism) and suggestive features (REM sleep behaviour disorder [RBD], neuroleptic sensitivity, and abnormal striatal dopaminergic imaging). The two main changes in the Fourth Consensus Report are (1) to upgrade RBD to become the fourth core clinical diagnostic feature and (2) to restructure the criteria so suggestive features no longer appear, but are replaced with "indicative biomarkers" and "supportive features."

2 | CORE FEATURES

RBD is a parasomnia in which movements and vocalisations occur during REM sleep (dream reenactments) because of the absence of normal REM atonia. The assessment toolkits recommend use of a specific validated question to identify RBD clinically. Where there is doubt about RBD, polysomnography (PSG) should be considered. The presence of two core clinical features is necessary to diagnose probable DLB whilst one alone enables a possible DLB diagnosis.

Less prominent than the upgrading of RBD, but helpful and important, is further clarification on parkinsonism. Whilst this has generally been understood to exclude drug-related and vascular parkinsonism, it has been less clear which and how many motor features of PD are required. PD requires the presence of bradykinesia (slowness of movement and decrement in amplitude or speed) together with rest tremor or rigidity or both.[3] The Fourth Report specifies that for counting as a core clinical feature for DLB, only one of these three features is sufficient. Special care is necessary when assessing older people or those with comorbidities, eg, osteoarthritis, or with advanced dementia because these features may be misinterpreted in such situations. For example, stiffness due to arthritis, or apraxia related to cognitive impairment, may mimic bradykinesia. In such situations, a dopaminergic scan should be considered. This leads to the other noticeable change in these revised diagnostic criteria, namely, the emphasis on biomarkers.

3 | INDICATIVE BIOMARKERS

In the previous Third Consensus Report, low dopamine uptake in the striatum on dopaminergic imaging was a suggestive feature of DLB. In the Fourth Report, this is joined (under the new category of indicative biomarkers) by abnormal (low uptake) cardiac MIBG (123-iodine-MIBG myocardial scintigraphy) imaging and PSG evidence of REM sleep without atonia. Abnormal MIBG imaging results from the reduction in noradrenergic innervation of the myocardium in Lewy body diseases[4] whilst PSG demonstrating REM sleep without atonia is the validated standard test for RBD.[5] The presence of any one of these in someone with dementia together with a core feature allows the diagnosis of probable DLB. Abnormal biomarker evidence, even more than one, in the absence of a core clinical feature only enables a possible DLB diagnosis. Those familiar with the Third Report will

notice that of the three suggestive features in those criteria (abnormal dopaminergic imaging, RBD, and severe neuroleptic sensitivity), neuroleptic sensitivity has been "downgraded" to a supportive clinical feature as "severe sensitivity to antipsychotic agents." This may be regarded as a good thing in that it reflects the greatly reduced use of antipsychotics in people with dementia generally and in those likely to have DLB in particular, with recent research reporting no study subjects having this feature (eg, Walker et al[6] and Donaghy et al[7]).

We have amended the toolkits to align with the new DLB criteria, to maximise ease of use and utility. Clinicians experienced in the diagnosis of DLB may not need to routinely use these toolkits for all patients, but our earlier study[1] found clinicians greatly valued the detail these toolkits provided about how to efficiently elicit the key features of DLB in everyday clinical practice. This was especially true for less experienced or trainee clinicians, and their routine use should serve as a useful training experience to heighten awareness of DLB symptoms and how to apply the new DLB diagnostic criteria.

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APPENDICES

ASSESSMENT TOOLKITS

Appendix 1 | Assessment Toolkit for Dementia with Lewy Bodies

Assessment Toolkit for Dementia with Lewy Bodies

Name:	Date of testing:
Date of birth:	Tester's name:
NHS No:	Informant:

Please use this Assessment toolkit in all people with cognitive decline. Below are the diagnostic features of dementia with Lewy bodies (DLB) at two levels of confidence (probable DLB and possible DLB) and on the following pages are specific questions to assist in the identification of the core and suggestive features of DLB.

DLB Diagnostic Criteria		Tick
1	Clinician diagnosis of dementia (cognitive decline sufficient to interfere with social/occupational function).	
2	Use screening questions below to cover the four domains of: cognitive fluctuation, visual hallucinations, RBD and parkinsonism.	
	Using your experience identify how many core and biomarker features of DLB are present (see below):	
3	**Core clinical features**	
	• Fluctuation in cognition	
	• Recurrent visual hallucinations	
	• REM sleep behaviour disorder	
	• One or more features of spontaneous parkinsonism	
4	**Indicative Biomarkers**	
	• Dopaminergic abnormalities in basal ganglia on SPECT/PET	
	• Low uptake on MIBG myocardial scintigraphy	
	• Polysomnography (PSG) confirmation of REM sleep without atonia	

Diagnose **Probable DLB** if either 2 core features are identified or 1 core and 1 indicative biomarker feature.	
Diagnose **Possible DLB** if any one feature is present. In such circumstances consider whether to refer subject for a dopaminergic SPECT scan (DaTSCAN), or MIBG or PSG, depending on local availability.	

Other Diagnoses	
Parkinson's Disease Dementia (PDD) (PD >1 yr before cognitive symptoms)	
Alzheimer's Disease	
Other Dementia	
MCI	

Patient informed of diagnosis.	**Yes**		**No**	

Questions to Identify Symptoms of DLB

Please respond to each of the questions below, asking carer or patient as appropriate.

Cognitive Fluctuation (to carer)

If two or more of these are answered 'Yes' the subject is highly likely to have cognitive fluctuation

1	Does the patient show moderate changes in their level of functioning during the day?	**Yes**		**No**	
2	Between getting up in the morning and going to bed at night, does the patient spend more than one hour sleeping?	**Yes**		**No**	
3	Is the patient drowsy and lethargic for more than one hour during the day, despite getting their usual amount of sleep the night before?	**Yes**		**No**	
4	Is it moderately difficult to arouse the patient so they maintain attention through the day?	**Yes**		**No**	

REM Sleep Disorder
(to carer = bed partner)

Have you ever seen the patient appear to ''act out his/her dreams'' while sleeping (punched or flailed arms in the air, shouted or screamed)?	**Yes**		**No**	

If answered affirmatively, then RBD is highly likely to be present.

REM Sleep Disorder
(to patient only if no bed partner and they have sufficient cognitive ability to be confident their answer is reliable)

Have you ever been told that you seem to "act out your dreams" while sleeping (punched or flailed arms in the air, shouted or screamed)?	**Yes**		**No**	

Visual Hallucinations

For the participant: Some people see things that other people cannot see.

1	Do you feel like your eyes ever play tricks on you?	**Yes**		**No**	
2	Have you ever seen something (or things) that other people could not see?	**Yes**		**No**	

For the carer:

1	Does the patient have hallucinations such as seeing false visions?	**Yes**		**No**	
2	Does he / she seem to see things that are not present?	**Yes**		**No**	

If, according to clinical judgement, visual hallucinations are present, determine as far as possible their frequency and recurrence. As a guide, visual hallucinations associated with DLB should not only occur during delirium, and are often recurrent over a period of months.

Assessment of Parkinsonism (5-item UPDRS)

Parkinsonism in DLB requires the presence of at least one of bradykinesia, rest tremor or rigidity. The 5-item UPDRS is a brief and validated scale for identifying parkinsonism in DLB (See below for further details)

POSTURAL TREMOR OF THE HANDS		
Normal	No tremor.	0
Slight	Tremor is present but less than 1 cm in amplitude.	1
Mild	Tremor is at least 1 but less than 3 cm in amplitude.	2
Moderate	Tremor is at least 3 but less than 10 cm in amplitude.	3
Severe	Tremor is at least 10 cm in amplitude.	4
KINETIC TREMOR OF THE HANDS		
Normal	No tremor.	0
Slight	Tremor is present but less than 1 cm in amplitude.	1
Mild	Tremor is at least 1 but less than 3 cm in amplitude.	2
Moderate	Tremor is at least 3 but less than 10 cm in amplitude.	3
Severe	Tremor is at least 10 cm in amplitude.	4
FACIAL EXPRESSION		
Normal	Normal facial expression.	0
Slight	Minimal masked facies manifested only by decreased frequency of blinking.	1
Mild	In addition to decreased eye-blink frequency, masked facies present in the lower face as well, namely fewer movements around the mouth, such as less spontaneous smiling, but lips not parted.	2
Moderate	Masked facies with lips parted some of the time when the mouth is at rest.	3
Severe	Masked facies with lips parted most of the time when the mouth is at rest.	4
GLOBAL SPONTANEITY OF MOVEMENT (BODY BRADYKINESIA)		
Normal	No problems.	0
Slight	Slight global slowness and poverty of spontaneous movements.	1
Mild	Mild global slowness and poverty of spontaneous movements.	2
Moderate	Moderate global slowness and poverty of spontaneous movements.	3
Severe	Severe global slowness and poverty of spontaneous movements.	4
RIGIDITY		
Normal	No rigidity.	0
Slight	Rigidity only detected with activation manoeuvre.	1
Mild	Rigidity detected without the activation manoeuvre, but full range of motion is easily achieved.	2
Moderate	Rigidity detected without the activation manoeuvre; full range of motion is achieved with effort.	3
Severe	Rigidity detected without the activation manoeuvre and full range of motion not achieved.	4

Total 5-item UPDRS Score =				
Is Parkinsonism present? (Use clinical judgement but for guidance a score >7 suggests significant parkinsonism is present, though a high score (>2) in a single domain may be sufficient to meet criteria)	**Yes**		**No**	

Appendix: Instructions for Assessing Parkinsonism (from UPDRS)

POSTURAL TREMOR OF THE HANDS

Rate each hand separately. Rate the highest amplitude seen. Instruct the patient to stretch the arms out in front of the body with palms down. The wrist should be straight and the fingers comfortably separated so that they do not touch each other. Observe this posture for 10 seconds.

KINETIC TREMOR OF THE HANDS

This is tested by the finger-to-nose maneuver. With the arm starting from the outstretched position, have the patient perform at least three finger-to-nose maneuvers with each hand reaching as far as possible to touch the examiner's finger. The finger-to-nose maneuver should be performed slowly enough not to hide any tremor that could occur with very fast arm movements. Repeat with the other hand, rating each hand separately. The tremor can be present throughout the movement or as the tremor reaches either target (nose or finger). Rate the highest amplitude seen.

FACIAL EXPRESSION

Observe the patient sitting at rest for 10 seconds, without talking and also while talking. Observe eye-blink frequency, masked facies or loss of facial expression, spontaneous smiling and parting of lips.

GLOBAL SPONTANEITY OF MOVEMENT (BODY BRADYKINESIA)

This global rating combines all observations on slowness, hesitancy, and small amplitude and poverty of movement in general, including a reduction of gesturing and of crossing the legs. This assessment is based on the examiner's global impression after observing for spontaneous gestures while sitting, and the nature of arising and walking.

RIGIDITY

Instructions to examiner: Rigidity is judged on slow passive movement of major joints with the patient in a relaxed position and the examiner manipulating the limbs and neck. First, test without an activation maneuver. Test and rate neck and each limb separately. For arms, test the wrist and elbow joints simultaneously. For legs, test the hip and knee joints simultaneously. If no rigidity is detected, use an activation maneuver such as tapping fingers, fist opening/closing, or heel tapping in a limb not being tested. Explain to the patient to go as limp as possible as you test for rigidity.

Appendix 2 | Assessment Toolkits for Lewy Body Dementia

Assessment Toolkits for Lewy Body Dementia

There are two toolkits, depending on whether the patient is presenting with a primary cognitive problem or with cognitive decline in the context of established Parkinson's disease.

One toolkit is for assisting in the diagnosis of Parkinson's Disease Dementia

This is therefore recommended for people with cognitive decline who have established Parkinson's disease (diagnosis for more than one year before the cognitive problems began).

The other toolkit is for assisting in the diagnosis of Dementia with Lewy Bodies

This toolkit is designed for use with people whose primary presenting problem is cognitive decline and who may or may not have evidence of recent Parkinson's disease (parkinsonian symptoms beginning at the same time or within a year of the cognitive symptoms).

Assessment Toolkit for Parkinson's Disease Dementia

Name:	Date of testing:
Date of birth:	Tester's name:
NHS No:	Informant:

Step 1: Please ask the following questions to the patient and/or his/her informant/carer:

Memory						Tick
Please ask the following questions about memory.						
1	Do you/does your relative have problems remembering things, e.g. what happened yesterday or what you were doing earlier?	Yes		No		
2	Do you/does your relative have difficulty remembering names of people you know well?	Yes		No		
3	When talking to people do you/does your relative often forget what had been said?	Yes		No		

Executive Impairment/Function						Tick
Please try to determine whether any difficulty is due to memory decline or physical impairment:						
1	Do you/does your relative have problems handling money or bank cards when paying for things?	Yes		No		
2	Do you/does your relative have difficulty looking after your/their own tablets?	Yes		No		
3	Are you/is your relative able to use household appliances on your own that you have used for a long-time, e.g. the TV or washing machine?	Yes		No		

Step 2: If Yes to 1 or more questions on memory AND 1 or more questions on executive impairment/function in step 1 then please administer the MOCA (or any other preferred cognitive assessment instrument to more fully assess for cognitive impairment).

Step 3: If MOCA<26 (or below cut-off for other instrument) and problems with everyday activities are due to memory decline and not due to physical impairment then please discuss with patient and/or carer/relative.

1	Seek confirmation of memory decline and related impairments in daily living activity.	**Yes**		**No**	
2	Ask how long have these memory problems been present: Have they been present for >1 year before Parkinson's disease	**Yes**		**No**	
3	Did these changes or difficulties develop gradually or rather than coming on suddenly?	**Yes**		**No**	
4	Do you think there was anything specific that caused these memory problems?	**Yes**		**No**	

Step 4: Now determine if the patient meets each of the 8 criteria below:

1	Clinician diagnosis of Parkinson's Disease.	
2	Onset of cognitive decline >1 year after onset of Parkinson's disease.	
3	Represents a decline from premorbid level.	
4	Deficits are severe enough to impair daily life (social, occupational, or personal care), independent of the impairment due to motor or autonomic symptoms.	
5	MOCA <21 or impaired on other cognitive test (if MOCA <26, diagnose PD-MCI if impairments in daily living are mild).	
6	A dementia syndrome with insidious onset and slow progression, developing within the context of established Parkinson's disease and diagnosed by history, clinical, and mental examination, defined as:	
	• Impairment in **more than one** cognitive domain from the MOCA:	
	• Attention: Serial 7s	
	• Executive functions: Lexical fluency, trails	
	• Visuo-spatial functions: Clock drawing, wire cube	
	• Memory: Recall of 5 objects.	
7	Absence of delirium, depression, systemic illness or drug intoxication sufficient to the cause cognitive impairment.	
8	Absence of other plausible cause of dementia, especially severe cerebrovascular disease.	

Please go to page 6 to confirm your clinical diagnosis.

Assessment Toolkit for Dementia with Lewy Bodies

Name:	Date of testing:
Date of birth:	Tester's name:
NHS No:	Informant:

Please use this Assessment toolkit in all people with cognitive decline. Below are the diagnostic features of dementia with Lewy bodies (DLB) at two levels of confidence (probable DLB and possible DLB) and on the following pages are specific questions to assist in the identification of core and suggestive features of DLB.

DLB Diagnostic Criteria		Tick
1	Clinician diagnosis of dementia (cognitive decline sufficient to interfere with social/occupational function).	☐
2	Use screening questions below to cover the four domains of: cognitive fluctuation, visual hallucinations, RBD and parkinsonism.	
	Using your experience to identify how many core and biomarker features of DLB are present (see below and next page):	
3	**Core clinical features** • Fluctuation in cognition • Recurrent visual hallucinations • REM sleep behaviour disorder • One or more features of spontaneous parkinsonism	☐ ☐ ☐ ☐
4	**Indicative Biomarkers** • Dopaminergic abnormalities in basal ganglia on SPECT/PET • Low uptake on MIBG myocardial scintigraphy • Polysomnography (PSG) confirmation of REM sleep without atonia	☐ ☐ ☐

Diagnose **Probable DLB** if either 2 core features are identified or 1 core and 1 indicative biomarker feature.	☐
Diagnose **Possible DLB** if any one feature is present. In such circumstances consider whether to refer subject for a dopaminergic SPECT scan (DaTSCAN), or MIBG or PSG, depending on local availability.	☐

Please go to page 6 to confirm your clinical diagnosis.

Questions to Identify Symptoms of DLB			Tick		
Please respond to each of the questions below, asking carer or patient as appropriate.					
Cognitive Fluctuation (to carer)					
If two or more of these are answered 'Yes' the subject is highly likely to have cognitive fluctuation					
1	Does the patient show moderate changes in their level of functioning during the day?	**Yes**		**No**	
2	Between getting up in the morning and going to bed at night, does the patient spend more than one hour sleeping?	**Yes**		**No**	
3	Is the patient drowsy and lethargic for more than one hour during the day, despite getting their usual amount of sleep the night before?	**Yes**		**No**	
4	Is it moderately difficult to arouse the patient so they maintain attention through the day?	**Yes**		**No**	

REM Sleep Disorder (to carer = bed partner)			
Have you ever seen the patient appear to ''act out his/her dreams'' while sleeping (punched or flailed arms in the air, shouted or screamed)?	**Yes**	**No**	
If answered affirmatively, then RBD is highly likely to be present.			

REM Sleep Disorder (to patient only if no bed partner and they have sufficient cognitive ability to be confident their answer is reliable)			
Have you ever been told that you seem to "act out your dreams" while sleeping (punched or flailed arms in the air, shouted or screamed)?	**Yes**	**No**	

Visual Hallucinations					
For the participant: Some people see things that other people cannot see.					
1	Do you feel like your eyes ever play tricks on you?	**Yes**		**No**	
2	Have you ever seen something (or things) that other people could not see?	**Yes**		**No**	
For the carer:					
1	Does the patient have hallucinations such as seeing false visions?	**Yes**		**No**	
2	Does he / she seem to see things that are not present?	**Yes**		**No**	

If, according to clinical judgement, visual hallucinations are present, determine as far as possible their frequency and recurrence. As a guide, visual hallucinations associated with DLB should not only occur during delirium, and are often recurrent over a period of months.

What is your clinical diagnosis?	Tick
Parkinson's Disease Dementia	
Parkinson's Disease MCI	
Parkinson's Disease	
Probable DLB	
Possible DLB	
Alzheimer's Disease	
Other Dementia	

	Tick
Patient Informed of Diagnosis?	