

# Management Guideline for Lewy body dementia



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# Management guideline development

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## How to reference this guideline

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## Part 1 Background

### Development of this Guideline

This guideline document has been produced as part of the NIHR Programme Grant funded DIAMOND-Lewy study [DTC-RPPG-0311-12001] entitled “Improving the diagnosis and management of dementias of Lewy body type in the NHS (DIAMOND-Lewy)”. The document offers practical advice to clinicians providing treatment and care to people with dementia with Lewy bodies (DLB) and Parkinson’s disease dementia (PDD) – collectively referred to as Lewy body dementia (LBD). It is complementary to our DLB and PDD diagnostic toolkits which are available, at <https://research.ncl.ac.uk/diamondlewy/publications/>.

In brief, we implemented a process-led approach in developing the LBD guidelines:

### *Establishment of the existing evidence base in LBD*

Formal systematic reviews and meta-analysis were conducted to capture all available, recent published information about the pharmacological and non-pharmacological management of LBD.[1-4] The level of evidence and grade of recommendations for each management strategy was assessed using the Oxford Centre for Evidence-Based Medicine criteria.[5] An important conclusion of the systematic reviews was that there is limited high-level evidence relating to pharmacological and non-pharmacological approaches, in meeting the needs of people who have LBD and their carers/families.

### *Public-patient workshops*

Two public-patient workshops were held with the participation of 38 people with LBD and their family/care-givers. The first event focussed upon identifying good practice in LBD clinical management, based on personal experiences. The emergent themes were developed in the second event and refined into a set of guiding principles and some specific statements. These were incorporated into the draft guideline that was circulated to the independent expert panel in a Delphi process.

Key themes are described below and should help the clinical team to deliver the guideline recommendations in a user-friendly way:

- It is important to recognise the carer’s expertise in knowing the person with dementia, and they need to feel that this is recognised. Whilst people want to be told what the clinician believes would be best, participants also expressed concern about doctors ignoring patient and carer preferences and not seeking help from other healthcare professionals who may be expert in certain symptoms. Carer diaries were mentioned as a useful aid to communicating needs. Opinions were divided as to whether or not patient and carer should be seen together by the clinical team, separately, or offered both. Some issues like increased care-giver stress, which are frequent in families and carer-givers dealing with LBD, probably needs to be explicitly discussed with the carer on their own.
- Although the advantages of developing and working through a problem list were recognised, it was also said that a long list of problems could be demoralising. This is particularly relevant to LBD where there are often many symptoms, not all of which may need to be formulated as problems. Reassurance that these symptoms can be expected as part of the disorder and not part of another illness is considered helpful.

- Carer and peer support groups are very valuable for support and practical help, providing a means of communication between people who have similar conditions and a source of practical information at an early stage on important general matters including, for example, Power of Attorney and financial advice and support (e.g. in the UK possible assistance with attendance allowance and council tax). Where geographical distance is a problem, phone or email contact can still be helpful.
- A clear message throughout the public-patient workshops was that although they appreciated the levels of care and concern that clinical teams offered, they often had difficulty in obtaining and understanding information about LBD. A lack of post-diagnosis support, advice and counselling was frequently mentioned, as was a shortage of easily accessible materials. As a result the guideline contains links to what were judged to be the best quality information sources at the time of writing and it is hoped that these will be made freely available to those attending our NHS services that will, in turn, be replaced by improved versions, adapted for local use.

#### *Drafting of guidelines and Delphi consensus*

Using the systematic reviews and public-patient feedback an initial draft of the guidelines was developed by the management guideline development team (J-P.T and IGM). Specific statements, framed under symptom domains, were created and submitted to an online anonymised online platform for review by our Delphi expert panel. The panel comprised 26 national and international experts from a range of disciplines (e.g. psychology, geriatrics, psychiatry, neurology, primary care, physiotherapy, nursing, and academic experts) identified through consultation with relevant stakeholder groups and supported by an extensive search of the literature for their publications, or their role as keynote speakers on management of LBD at, for example, major conferences. The Delphi process was conducted over three rounds. A high level of agreement was sought across the three rounds (85% for rounds 1 and 2 and 75% for round 3). Controversial statements were modified on the basis of feedback and rerun in the subsequent round or removed. Of 252 original statements, 161 were kept, with 78 of these (48.4%) gaining full consensus panel agreement for inclusion, 52 (32.3%) with 90% to 99% consensus agreement, and 31 statements (19.3%) agreed by 75% to 89% of the panel. After this process, the guideline statements were re-collated and formulated into one document. More controversial statements (but still meeting majority consensus opinion > 50%) or points of clarification are included as footnotes in the guideline.

From the reference guideline, we generated one-page summaries of symptom management (cognitive, neuropsychiatric, motor, autonomic and sleep) as well as an overview summary page. These resources are intended to be used as aide-mémoires, as print-outs for use in clinic, or as the basis of developing specific local care pathways for LBD. The guideline and one page summaries together form our LBD management toolkit.

#### *Piloting in a clinical study*

The draft management toolkit (version 1.0) was successfully piloted in one NHS service where staff and patients from Old Age Psychiatry and Parkinson's disease services were engaged. Their feedback was used to revise and modify the guideline and one page summaries (version 2.0). This management toolkit was subsequently used in a multi-centre cluster based randomised control trial as part of the NIHR DIAMOND-Lewy study.

### *Updates to the guideline*

The current management guideline is a living document and will be periodically updated to reflect changes in clinical practice and new treatments as they come available. An update history is provided below:

**October 2018:** A further literature review (up to, and including October 2018) was conducted to identify relevant and new pharmacological and non-pharmacological therapies in LBD. Additionally we asked our expert panel group to identify any new therapeutic approaches applicable to the management guidelines. From these processes, 30 new or modified management statements were drafted and re-run using the same Delphi consensus process described above with the acceptance of 26 of 30 of these statements. These were subsequently added to the revised guidelines (version 3.0).

**September 2019:** Complimentary to this guideline we have published a comprehensive review on the management of LBD which provides context and background to the guideline and summary sheets:

Taylor, J-P. et al. *New evidence on the management of Lewy body dementia*. The Lancet Neurology [http://dx.doi.org/10.1016/S1474-4422\(19\)30153-X](http://dx.doi.org/10.1016/S1474-4422(19)30153-X)



## Part 2 How to use this document

Management of LBD is largely initiated in NHS secondary care and specialist settings typically involving multiple clinical services including old age psychiatry, neurology, and care of the elderly. Monitoring, support and continuing care of patients and families is then often shared with primary care services, social care and voluntary sector organisations.

Detailed guidance is available from NICE (NG97) about general principles of good practice in supporting people with dementia and their carers in health and social care. It includes recommendations for people with DLB but not for PDD, as PDD is covered in the NICE Parkinson's disease guideline (NG71). This guideline is intended to be used in the context of the NICE recommendations, as a resource to offer more detailed information about clinical care and management of DLB and PDD. We assume that users will already be addressing the important general dementia needs of their LBD patients and carers including issues such as consent and advance decision making; managing comorbidities, maximising functional abilities and independence, assessing competence to drive and to manage finances; palliative care and providing support for family members and carers at all stages.

**Within the guideline, consensus agreed management statements are indicated by a ✓**

As noted previously, footnotes in the guideline indicate either clarification points or selected comments made by Delphi panellists in response to the management recommendations and which do not necessarily represent the consensus view (but still meeting a majority consensus opinion > 50%).

### References

1. Stinton, C., et al., *Pharmacological Management of Lewy Body Dementia: A Systematic Review and Meta-Analysis*. Am J Psychiatry, 2015: p. appiajp201514121582.
2. Wang, H.F., et al., *Efficacy and safety of cholinesterase inhibitors and memantine in cognitive impairment in Parkinson's disease, Parkinson's disease dementia, and dementia with Lewy bodies: systematic review with meta-analysis and trial sequential analysis*. J Neurol Neurosurg Psychiatry, 2015. **86**(2): p. 135-43.
3. Connors, M.H., et al., *Non-pharmacological interventions for Lewy body dementia: a systematic review*. Psychol Med, 2017: p. 1-10.
4. Morrin, H., et al., *Systematic review of the efficacy of non-pharmacological interventions in people with Lewy body dementia*. Int Psychogeriatr, 2018. **30**(3): p. 395-407.
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## Part 3 Basic principles of LBD management

### Principles of management - general recommendations

- ✓ Management plans should be discussed after a diagnosis has been given, with the opportunity for questions and clarification from patient and family/carers.
- ✓ Identify key problems under domain headings such as cognition; gait, balance and movement; hallucinations; fluctuations; behaviour and mood; sleep, and autonomic system dysfunction.<sup>1</sup>
- ✓ Establish which problems have high priority for treatment, taking into account the views of the patient, family/carers and the treating professional. It may be helpful to rank order the importance/severity of symptoms to aid the decision regarding the order in which to treat them.
- ✓ There should be a full discussion with the patient and carers about the possible benefits and risks of treatment.
- ✓ It is particularly important to explain that symptom response is variable and that benefits in one might be at the cost of worsening of others.
- ✓ Due to the inherent fluctuations in LBD it may not always be easy in the short term to directly link changes in symptoms, with treatment changes.<sup>2</sup>
- ✓ Introduce one treatment at a time and try to establish best dose and effectiveness before moving to additional treatments.
- ✓ Although individual treatments may be aimed at specific targets (e.g. cholinesterase inhibitors for cognitive impairments) be aware that they may also affect other domains<sup>3</sup> e.g. hallucinations or fluctuations or may be non-specific i.e. global.

### Use of medication

Polypharmacy is common in older people with dementia including LBD and many may be taking inappropriate medications which could contribute to symptoms such as sedation, cognitive impairment, falls, delirium and behavioural disturbances e.g. opiates, anticholinergics.

LBD patients may exhibit exaggerated responses to several types of medication, reflecting their multiple underlying neurotransmitter deficits.

This can be to the benefit of the patient, such as the often large clinical response to cholinesterase inhibitors or it can be detrimental. Of the latter, severe sensitivity to antipsychotic agents is the best documented, with both typical and atypical antipsychotics having potential to provoke a severe adverse reaction in up to 50% of patients with LBDs even when low doses are used. Severe antipsychotic sensitivity reactions include a sudden and profound worsening of parkinsonism accompanied by sedation, confusion, and autonomic dysfunction: symptoms not dissimilar to that of a neuroleptic malignant syndrome. Intensive support may be required to manage such reactions which have a high mortality rate if not recognised and dealt with.

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<sup>1</sup> In particular, consider the impact of these problems on patients and carers in terms of their quality of life and day to day function.

<sup>2</sup> Carers and family members are often best placed to pick up on subtle changes in symptoms.

<sup>3</sup> Be aware that what is a side effect with a treatment in one person may be a benefit in another patient. For example cholinesterase inhibitors may increase bowel movements; in one individual this may cause diarrhoea whereas in another, the increased gut motility may help with constipation.

## Use of medication - general recommendations

(Class specific recommendations are given separately)

- ✓ As part of good prescribing practice: check for drug-drug interactions; consider using compliance aids; minimising the number of prescribers and minimising polypharmacy where possible.
- ✓ Regularly review all psychotropic and physical health medications that a patient is currently taking.
- ✓ Review the need for common drugs which can affect brain function and/or cause sedation and falls e.g. opiates, antihistamines, anticonvulsants, benzodiazepines.
- ✓ Minimise anticholinergic burden<sup>4</sup> as this may worsen cognition and behaviour, and counteract cholinesterase inhibitors.
- ✓ Prominent warnings with regards to the use of antipsychotic medication should be added to patient (electronic and paper) notes and included in correspondence with other health professionals.
- ✓ If the patient is already on an anti-psychotic, ensure that it is not causing significant motor or cognitive side effects.

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<sup>4</sup> Anticholinergic cognitive burden scales can be helpful in this regard.

## Part 4 Managing cognitive symptoms

Although cognitive impairments will by definition be present in all LBD patients, they will vary in their severity, variability, and importance to the patient and carer compared with other symptoms present.

### Managing cognitive symptoms - general recommendations

- ✓ The treating clinician needs to establish the presence of significant cognitive difficulties warranting treatment. These may include impairments in cognition including memory, attention, fluctuating cognition, executive functioning, visuo-perceptual abilities, and disorganised speech/communication.
- ✓ Evidence for cognitive difficulties should be obtained from reports of the patient and an informed carer and the results of formal cognitive testing.<sup>5</sup>
- ✓ Cognitive fluctuations, whilst intrinsic to LBD, may also be a feature of provoked delirium. Therefore, exclusion of the latter is important. Other factors causing or aggravating cognitive decline should also be excluded.
- ✓ Non-pharmacological approaches to managing cognitive impairments include cognitive stimulation, use of memory aids, increased social interaction and stimulation, and exercise.<sup>6</sup>

### Medications

#### *Cholinesterase inhibitors*

##### General principles

- ✓ The first line treatment of cognitive impairment is with cholinesterase inhibitors.
- ✓ Clinician choice will be influenced by previous experience, ease of administration, dose titration regime and side effect profile.
- ✓ Systematic reviews have shown evidence that donepezil and rivastigmine are similarly effective in DLB and PDD for both cognitive and neuropsychiatric outcomes.
- ✓ Galantamine may have positive effects on cognition and neuropsychiatric symptoms but data are limited.
- ✓ Before starting cholinesterase inhibitors, enquire/examine for evidence of clinically significant cardiovascular disease, particularly orthostatic hypotension, syncope or pre-syncope or cardiac dysrhythmia / conduction disturbance or bradycardia.
- ✓ Consider carrying out an ECG before cholinesterase inhibitor, particularly if there is a history of cardiac issues and/or autonomic dysfunction.
- ✓ Cardiology referral should be made in cases of uncertainty including decisions regarding fitting of pacemakers.

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<sup>5</sup> Depending upon the clinical setting, it is recognised in some cases that this may not always be possible.

<sup>6</sup> Note the evidence base for these interventions is lacking, at present, in LBD although evidence of their efficacy more broadly in dementia is reasonably established.

## Dose and titration

- ✓ Cholinesterase inhibitors are best titrated to the maximum tolerated dose and maintained at this level.<sup>7,8</sup>
- ✓ Dose and titration for donepezil should proceed as follows: 5mg once daily for 4-6 weeks, increased to 10mg daily if no significant side effects occur.
- ✓ Dose and titration for rivastigmine should proceed as follows: 1.5 mg twice daily for 4 weeks, increased to 3 mg twice daily.<sup>9</sup> Dose can be increased up to 4.5 mg twice daily going up to 6 mg twice daily if no significant side effects occur.
- ✓ A rivastigmine patch is also available and this may have advantages where there are issues with swallowing difficulties, compliance or if there is a history of significant response variation to oral dosing. Dosing and titration is typically 4.6 mg/24 hours for 4 weeks, increased to 9.5 mg/24 hours with a further increase, after 4 weeks, to 13.3 mg/24hours if no significant side effects.<sup>10</sup>
- ✓ Dose and titration for galantamine should proceed as follows: 8mg/day increased to the initial maintenance dose of 16mg/day after a minimum of 4-6 weeks. A further increase of 24mg/day of galantamine can be attempted after 4 weeks at 16mg/day if no significant side effects occur.

## Assessing response and deciding about continuation

- ✓ Record baseline cognitive performance using a preferred scale.
- ✓ Global and behavioural / psychiatric baseline symptoms should also be documented.
- ✓ Assess outcome after 3-6 months on maximum tolerated dose (be aware that some patients may take longer to respond). Cognitive, global and other domain assessments may be used to support this.
- ✓ Once optimised, treatment should be continued for as long as the patient/carer/clinician consensus is that there are positive risk/benefits.
- ✓ Due to the progressive nature of LBD it is likely that measures will eventually fall below baseline levels but this alone should not be taken as lack of continuing response.
- ✓ If/when discontinued, cholinesterase inhibitors should be withdrawn gradually as there are reports of a rebound worsening of symptoms.
- ✓ Strategies for non-response or poor tolerance to one cholinesterase inhibitor include switching to another cholinesterase inhibitor, but there is no systematic evidence to support this approach.

## Adverse effects

- ✓ In addition to gastrointestinal symptoms, cholinesterase inhibitors may also cause or exacerbate the following:
  - postural hypotension
  - urinary frequency

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<sup>7</sup> Some patients may benefit from dose escalation beyond the normal range; however this is often likely to be accompanied by increasing side effects and there are no safety data available for use of cholinesterase inhibitors above the recommended maximum dose in LBD.

<sup>8</sup> Timing of administration may also be important. Once daily cholinesterase inhibitors are typically given in the day as they can worsen sleep if given in the evening. However if dizziness and nausea are present then changing administration of the cholinesterase inhibitor to the evening may help.

<sup>9</sup> Target dose of oral rivastigmine, if tolerated, should be 6 mg bd.

<sup>10</sup> Ensure that the patch is removed after 24 hours and switch skin site regularly.

- hyper-salivation
- watering eyes
- runny nose
- worsening of extrapyramidal motor symptoms, particularly fine tremor.

These adverse effects may improve with dose reduction.

## Memantine

### General principles

- ✓ Although data are limited and inconsistent, memantine may be used in LBD either:
  - as monotherapy if cholinesterase inhibitors are not tolerated, or, if there are any other contra-indications to the use of cholinesterase inhibitors, or,
  - in combination with cholinesterase inhibitors, particularly if the effectiveness of the cholinesterase inhibitor is limited or is declining, or the disease is becoming more severe.

### Dose and titration

- ✓ Dosing of memantine should be increased to 20mg once daily over 4 weeks according to tolerance as per the recommendations for treating patients with AD.<sup>11, 12</sup> Some patients may prefer divided dosing.

### Adverse effects

- ✓ Adverse effects of memantine include gastrointestinal symptoms, confusion, somnolence, hypertension and dizziness.
- ✓ Be cautious in prescribing memantine to individuals with a history of seizures, or poor renal function.
- ✓ Memantine may enhance the effects of dopaminergics and selegiline, as well as being toxic when given with amantadine.

### Assessing response and deciding about continuation

- ✓ Record baseline cognitive performance using a preferred scale.
- ✓ Global and behavioural / psychiatric baseline symptoms should also be documented.
- ✓ Assess outcome after 3-6 months on maximum tolerated dose (be aware that some patients may take longer to respond). Cognitive, global and other domain assessments may be used to support this.
- ✓ Once optimised, treatment should be continued for as long as the patient/carer/clinician consensus is that there are positive risk/benefits.
- ✓ Due to the progressive nature of LBD it is likely that measures will eventually fall below baseline levels but this alone should not be taken as a lack of continuing response.

<sup>11</sup> Start memantine at 5mg daily and increase by 5mg per week to maximum of 20mg daily.

<sup>12</sup> In patients with estimated glomerular filtration rate (eGFR) <50ml/min dose adjustments of memantine may be required.

## Part 5 Managing neuropsychiatric symptoms

Neuropsychiatric symptoms are common in LBD and include visual hallucinations, delusions, hallucinations in other modalities, agitation, aggression, apathy, depression and anxiety.

- ✓ The general approach to assessment and non-pharmacological management of such symptoms described in NICE guidelines may be helpful, although there are no specific recommendations as to how these might be adapted for people with LBD.
- ✓ Whilst a selected target symptom approach is generally to preferred, the individual symptoms are often closely linked, and the assessment of composite neuropsychiatric function may be more clinically meaningful.
- ✓ The presence of a symptom per se, does not necessarily mean that treatment is required as a priority, rather this should be offered in response to patient or carer distress or request.

### Managing neuropsychiatric symptoms - general recommendations

- ✓ The treating clinician needs to establish the presence, severity and impact of significant neuropsychiatric symptoms warranting treatment. These may include visual hallucinations, hallucinations in other modalities, delusions, apathy, agitation, aggression, depression and anxiety.
- ✓ Evidence for problems with neuropsychiatric symptoms should be obtained from reports of the patient and an informed carer; systematic rating scales may be helpful.
- ✓ Other factors causing or aggravating mood and behaviour disturbance should be excluded. These will include physical illness, pain, hunger or discomfort, delirium, and other environmental precipitants and perpetuating factors.

### Hallucinations and delusions

Visual hallucinations and related symptoms including illusions and misidentifications are common in LBD, often precipitating the initial clinical presentation, especially if the images are frightening or unpleasant in content.<sup>13</sup> Delusional beliefs in LBD are commonly secondary explanations of these experiences. In a minority the hallucinations may be regarded neutrally or sometimes even as comforting or pleasurable and the patient may not be seeking treatment.

### Hallucinations and delusions - treatment recommendations

- ✓ Simple explanation of visual symptoms as a consequence of impaired visual processing may allay fears and avoid the need for medication.
- ✓ Interventions such as removing cushions, patterned curtains and other stimuli that might precipitate visual misinterpretations can be helpful, as is provision of good lighting.
- ✓ Confronting and challenging the unreality of hallucinations and delusions by care-givers / family members is often not helpful. In some cases, distraction from the hallucinatory experience can be tried.
- ✓ If symptoms persist and are troublesome, a trial of medication may be required.
- ✓ People with LBD who have non-cognitive symptoms causing significant distress to the individual, or leading to behaviour that challenges, should be offered a cholinesterase inhibitor.<sup>14</sup>

<sup>13</sup> A formal ophthalmologic evaluation may be required for some in order to investigate for the possibility of primary ocular causes of any visual symptoms given the former are common in the older population.

<sup>14</sup> A second-line treatment may be memantine, although it should be noted that the reported efficacy of memantine on neuropsychiatric symptoms in patients with LBD is mixed.



- ✓ Cholinesterase inhibitor or memantine prescribing for non-cognitive symptoms should follow the guidelines described earlier for the treatment of cognitive symptoms, although the response may be judged earlier.
- ✓ If these are ineffective a trial of an antipsychotic agent may need to be considered.

#### Antipsychotic prescribing in LBD

- ✓ There should be a full discussion with the person with dementia and/or carers about the possible benefits and risks of antipsychotic treatment including neuroleptic sensitivity and the possible increased risk of stroke/transient ischaemic attack and possible adverse effects on cognition.<sup>15</sup>
- ✓ A key element in managing neuropsychiatric symptoms (particularly agitation, aggression and delusions and hallucinations not responsive to cholinesterase inhibitors) is extreme caution in the use of antipsychotic medications because of the risk of adverse effects including severe sensitivity reactions, and lack of documented efficacy.
- ✓ Target symptoms should be identified, quantified and documented and changes in these should be assessed and recorded at regular intervals.
- ✓ Changes in cognition should be assessed and recorded at regular intervals.
- ✓ Alternative medication should be considered if necessary.
- ✓ The effect of comorbid conditions, such as depression, should be considered
- ✓ The choice of antipsychotic should be made after an individual risk–benefit analysis.
- ✓ The lowest possible dose should be initiated and then titrated upwards.
- ✓ Treatment should be time limited and regularly reviewed (according to clinical need).
- ✓ The prescriber should monitor carefully for the emergence of severe antipsychotic sensitivity reactions.
- ✓ In the event of a suspected antipsychotic sensitivity reaction the drug should be withdrawn and the patient closely observed with view to providing active support should further deterioration occur.
- ✓ Clozapine, which is effective in PD psychosis, may also help in LBD, although evidence is lacking.<sup>16</sup> There is no evidence to favour any other antipsychotic drug in LBD<sup>17</sup> although quetiapine appears to have the least adverse effects.<sup>18</sup>

#### Agitation and aggression - treatment recommendations

- ✓ Agitation and aggression in a patient with LBD are likely to be multi-factorial and the clinician needs to identify the relevant antecedent and perpetuating factors, in order to manage these.
- ✓ Behavioural disturbance in LBD may sometimes be driven by hallucinatory and other psychotic symptoms and may improve when these are treated with a cholinesterase inhibitor and/or memantine.
- ✓ If anti-psychotics are used to manage agitation and aggression, particular caution needs to be taken as previously described.

<sup>15</sup> This discussion should be recorded in medical notes.

<sup>16</sup> Clozapine requires monitoring for blood dyscrasias and may cause other side effects in LBD; many of these relate to its anticholinergic properties.

<sup>17</sup> The 2017 Parkinson's Disease NICE guidelines (NG71) recommend that olanzapine is specifically avoided in Parkinson's disease for the treatment of delusions and hallucinations.

<sup>18</sup> Care should be taken with dose titration of quetiapine and the risk of orthostatic hypotension and increased sedation.



### Apathy - treatment recommendations

- ✓ Providing adequate environmental stimulation may help reduce apathy.<sup>19</sup>
- ✓ Apathy may also improve with a cholinesterase inhibitor.

### Depression and anxiety - treatment recommendations

- ✓ In the treatment of depression consideration should be given to use of social interventions to enhance mood.
- ✓ If antidepressant medication is given, drugs with significant anti-cholinergic side effects should be avoided.
- ✓ Evidence for antidepressant drug efficacy and tolerability in LBD is limited. Some trials have been positive for selective serotonin reuptake inhibitors, serotonin-norepinephrine reuptake inhibitors and nortriptyline<sup>20</sup> in Parkinson's disease.
- ✓ Whilst there is no evidence base, cholinesterase inhibitors may help some particularly if there is an apathy component.
- ✓ Electroconvulsive therapy can be considered for the acute treatment of severe depression in LBD that is life-threatening and when a rapid response is needed although there needs to be awareness that there may be associated side-effects e.g. confusion and autonomic dysfunction.
- ✓ There is insufficient evidence to support the use of transcranial magnetic stimulation in the treatment of depression in LBD.

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<sup>19</sup> Evidence for this appears to be anecdotal, however.

<sup>20</sup> Nortriptyline may have significant anti-cholinergic effects. Paroxetine (SSRI) is also associated with moderate anticholinergic effects.

## Part 6 Managing Parkinsonian symptoms

### General principles for the management of Parkinsonian symptoms in dementia with Lewy bodies

- ✓ Establish the presence of significant motor difficulties which are impairing function and warrant treatment. These may include significant resting tremor, marked bradykinesia, freezing episodes, gait disturbance, and/or rigidity.
- ✓ Identify other factors which may be a cause of a worsening of motor function such as concurrent cholinesterase inhibitor or antipsychotic use, locomotor problems secondary to osteoarthritis, increasing frailty etc.
- ✓ Be aware that parkinsonian symptoms may be less treatment-responsive in LBD than in PD.
- ✓ Physiotherapy has a role and clear evidence base in Parkinson's disease and may help with freezing of gait, gait re-education, improvement in balance, power and flexibility as well as enhance mobility, decrease the risk of falls and improve functional independence. In DLB (and PDD) cognitive impairment and other comorbid symptoms can influence engagement with therapy but outcomes may still be positive.
- ✓ Occupational therapy assessment and home adaptations can help reduce the impact of motor difficulties and reduce falls risk.
- ✓ Given increased falls risk in DLB (and PDD) vitamin D supplementation should be considered where appropriate.<sup>21</sup>

### Managing Parkinsonian symptoms in dementia with Lewy bodies - treatment recommendations.<sup>22</sup>

- ✓ The preferred pharmacological treatment of parkinsonism in LBD is levodopa monotherapy.
- ✓ Use the minimal levodopa dose required for benefit.
- ✓ Either co-careldopa or co-beneldopa may be used.
- ✓ Start low, and increase dose slowly: typical initiation doses are lower than in Parkinson's disease (e.g. 50 mg (expressed as levodopa dose) taken 1-3 times daily).<sup>23, 24</sup>
- ✓ Monitor closely for adverse effects including psychosis, sedation, postural hypotension, nausea and vomiting.
- ✓ Consider speech and language therapy referral for motor related speech and swallowing problems.

### General principles for the management of Parkinsonian symptoms in Parkinson's disease with dementia

The general principles are similar to those for DLB but PDD patients will usually already been on one or more anti-parkinsonian agents. Research suggests that although anti-parkinsonians are of themselves not generally sufficient to cause neuropsychiatric symptoms, they can do so in people with Lewy body disease, particularly those with dementia. Management decisions are therefore typically around dose reduction/cessation.

### Managing Parkinsonian symptoms in Parkinson's disease with dementia - treatment recommendations

- ✓ To avoid exacerbating neuropsychiatric problems without leading to unacceptable immobility, a gradual and systematic simplification of the anti-parkinsonian drug regimen is often necessary.

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<sup>21</sup> Vitamin D deficiency is more common in women and in the elderly.

<sup>22</sup> Zonisamide 25mg to 50 mg once a day as an adjunct to levodopa may have some motor benefits in PD and DLB.

<sup>23</sup> For example, this would equate to co-careldopa dose of 12.5mg carbidopa/50mg levodopa.

<sup>24</sup> Some patients may tolerate and respond to higher doses although titration needs to be done slowly.

- ✓ Where anti-parkinsonian drugs are being altered this should be done in close collaboration with the original prescriber of the medicines where possible.
- ✓ Withdraw anticholinergic drugs, amantadine, monoamine oxidase B inhibitors, dopamine agonists and catechol-O-methyltransferase inhibitors, one at a time.

## Part 7 Managing autonomic dysfunction

There are a wide range of autonomic symptoms in LBD including: orthostatic hypotension, gastroparesis, constipation, gastroparesis, urinary dysfunction, urinary incontinence, sexual dysfunction, excessive sweating, dysphagia and sialorrhoea.

However there is no established evidence base for the treatment of these symptoms and treatment is on the basis of clinical judgement.

### Orthostatic hypotension - general principles

- ✓ There is insufficient evidence to recommend any specific treatments for the management of orthostatic hypotension in LBD. However a number of broad principles apply. Non-pharmacological interventions are advised as first-line.
- ✓ Medications (e.g. levodopa, dopamine agonists, antihypertensives, antidepressants, alpha-adrenergic blockers, sildenafil)<sup>25</sup>, dehydration, cardiac disease, fever and anaemia may cause or exacerbate orthostatic hypotension.
- ✓ Orthostatic hypotension may manifest at particular times, for example at mealtimes, when taking alcohol, early morning, during defaecation or micturition, and with physical activity.
- ✓ If there is significant dizziness, falls or episodes of loss of consciousness, or symptomatic orthostatic hypotension consider a referral to a specialist falls/ syncope clinic.

### Orthostatic hypotension - treatment recommendations

#### Non-pharmacological management strategies

- ✓ Advise the patient to stand slowly.
- ✓ Slight increases in salt intake may help some.<sup>26</sup>
- ✓ The use of compression hosiery.<sup>27</sup>
- ✓ Raising the head of the bed by 10-15 cm may help some, particularly those with orthostatic hypotension in the early morning.
- ✓ Increase fluid intake – usually 2 litres in total, daily, is advised.<sup>28</sup>

#### Pharmacological management strategies

Examples of agents<sup>29</sup> that may be tried include:

- ✓ Fludrocortisone (50-300 mcg / day). This drug is a potent mineralocorticoid and can have significant dose related adverse effects including electrolyte disturbances, hypertension and oedema. Titrate slowly and monitor electrolytes (particularly potassium).<sup>30</sup>

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<sup>25</sup> Be aware that cholinesterase inhibitors can also be a contributor.

<sup>26</sup> Consider supplemental sodium (e.g. 1-2g three times a day) if urinary sodium is less than 170 mmol/24 hours.

<sup>27</sup> May not suitable if there is significant peripheral vascular disease.

<sup>28</sup> Be cautious with fluid amount in older patients and/or presence of heart failure. Taking fluid as a bolus has been advocated by some.

<sup>29</sup> Caffeinated drinks may help increase BP in some. Droxidopa (initially 100 mg three times daily up to maximum of 600 mg three times daily) has been used for treatment of orthostatic hypotension in United States but is not licensed in the United Kingdom.

<sup>30</sup> It is advised that patients should carry a steroid treatment card if on fludrocortisone.

- ✓ Midodrine (2.5 – 10 mg up to three times a day). There is also a need to regularly monitor blood pressure due to the risk of supine hypertension as well as hepatic and renal function.<sup>31</sup> Avoid evening doses of midodrine; last dose should be taken at least four hours before bed.<sup>32</sup>
- ✓ Be aware that some medications for orthostatic hypotension may cause severe supine hypertension.

#### Gastroparesis- treatment recommendations

- ✓ Thickened fluids, carbonated liquids and sugar free chewing gum may help.
- ✓ Be aware that dopaminergic medications can exacerbate gastroparesis.<sup>33</sup>
- ✓ Advise the patient to have small and frequent meals and drink during meals. Avoidance of high fat foods may also help as well as walking after meals.
- ✓ Domperidone (10 mg up to 3 times per day) may help some patients with gastroparesis but there are significant concerns with regard to cardiotoxicity and the risk of QT prolongation.<sup>34</sup>
- ✓ Avoid using metoclopramide given its central dopamine antagonist effect.
- ✓ Giving levodopa in solution may help with patients with significant motor fluctuations and delayed gastric emptying.
- ✓ Alternatively, for some patients with delayed gastric emptying, their motor fluctuations may be improved through jejunal administration of levodopa.

#### Constipation - treatment recommendations

- ✓ Constipation can be caused directly by the disease process and can also be caused by a variety of factors including poor fluid intake, reduced fibre intake, sedentary behaviour and medication use (e.g. opiates and some anti-parkinsonian drugs). Tackling these factors may help the constipation.<sup>35</sup>
- ✓ Check there has been no significant changes in bowel habits (such as PR bleeding, weight loss and anaemia) which may indicate a more sinister cause e.g. colorectal malignancy.
- ✓ Be aware that outlet constipation can be caused by dystonic anismus; specialist advice may need to be sought for the assessment and management of this.
- ✓ Conservative measures should be considered first line such as increasing levels of daily activity, eating more fruit and vegetables and increasing fluid intake.<sup>36</sup>
- ✓ Short courses of bulk forming / osmotic laxatives, e.g. macrogol, may help constipation in some.
- ✓ Mild stimulant laxatives can be used, if required<sup>37</sup>. Typical agents might include senna (7.5-15mg at night), bisacodyl (5-10 mg at night) or sodium docusate (50 to 400 mg in divided doses each day).<sup>38</sup>
- ✓ Lubiprostone may be a useful second line treatment option.

<sup>31</sup> Needs specialist to initiate.

<sup>32</sup> Do not give if there is a suspicion of significant post void residual urine.

<sup>33</sup> Conversely, some experts felt that cholinesterase inhibitors may help with gastroparesis.

<sup>34</sup> If risk of QTc prolongation will need ECG before starting and after one week of treatment. If prescribed longer term will need regular review.

<sup>35</sup> Effect of treatment can be monitored using stool charts e.g. Bristol Bowel chart.

<sup>36</sup> Probiotics have been suggested to help improve the number of completed bowel movements in people with Parkinson's disease.

<sup>37</sup> Type of laxative used may be influenced by stool type

<sup>38</sup> If these are not helping, stronger laxatives, suppositories or enemas may need to be considered; however seek specialist advice in these circumstances.

- ✓ Stool softeners can be helpful if stools are very hard.<sup>39</sup>
- ✓ Mild suppositories such as glycerine may help also bowel emptying.

#### Urinary dysfunction and incontinence - treatment recommendations

- ✓ It is important to clarify the potential cause of any urinary dysfunction as this will influence treatment. For example, per rectum examination (prostate enlargement) and bladder scans to ensure post void residuals < 100ml may be useful.
- ✓ Exclude constipation or excess caffeine as potential causes/exacerbating factors.
- ✓ Non-pharmacological strategies should be employed first line.
- ✓ Regular, prompted, voiding with use of incontinence pads may be helpful.
- ✓ Consider referral to an incontinence nurse and / or urology if symptoms are particularly troublesome or have never been previously investigated.
- ✓ Avoidance or reduction in diuretics may help if there are no contraindications.
- ✓ Be aware that cholinesterase inhibitors can precipitate urgency and urge incontinence.
- ✓ Bladder anticholinergics should be avoided, in particular, the use of agents which have a significant centrally acting effect such as oxybutynin and tolterodine.<sup>40</sup>
- ✓ Mirabegron may be an alternative to anticholinergics for bladder overactivity.<sup>41</sup> Typical dose is 25 mg per day although may be increased to 50 mg per day depending upon efficacy and tolerability.
- ✓ Intravesical botulium toxin may have a positive effect on neurogenic detrusor overactivity in those intolerant of anticholinergics.<sup>42</sup>

#### Male sexual dysfunction - treatment recommendations

- ✓ In men, the use of phosphodiesterase-5 inhibitors such as sildenafil can be considered for erectile dysfunction. However, these need to be prescribed with caution if the patient has postural / orthostatic hypotension.<sup>43, 44</sup>

#### Excessive sweating - treatment recommendations

- ✓ General advice to patients includes:
  - ✓ Wear loose fitting, natural fibre clothing and use natural light cotton bedding if there are significant night sweats.
  - ✓ If practical, to frequently shower and change clothes.
  - ✓ Use of moisture-absorbent socks and underarm dress shields may help absorb excess sweat.
  - ✓ Antiperspirants can help some.

<sup>39</sup> Bulking agents such as bran or ispaghula husk may be options but require a fluid intake that people with LBD can find hard to maintain.

<sup>40</sup> Newer, larger and less lipophilic compounds such as trospium may have less propensity to cross BBB and cause cognitive side effects.

<sup>41</sup> Use with caution in renal or hepatic impairment; use of mirabegron is contraindicated if impairment is severe. Caution is also required if there is a history of Q-T prolongation and its use should be avoided if there is severe uncontrolled hypertension.

<sup>42</sup> Urinary retention is a potential risk with this treatment and patients may need to self-catheterise.

<sup>43</sup> Be aware that depression and anxiety, common in LBD can cause problems with erectile dysfunction. In addition certain medications can exacerbate e.g. serotonin re-uptake inhibitors.

<sup>44</sup> Referral to a specialist service/urologist may help some especially if other interventions need to be considered e.g. intra-urethral prostaglandin, prostaglandin injection, vacuum pumps etc.

- ✓ Avoid foods and situations which trigger sweating e.g. alcohol, spicy foods, hot rooms etc.
- ✓ Ensure adequate fluid intake to replace losses.
- ✓ Excessive sweating may be exacerbated by any dopamine replacement therapy and can occur during “off” periods. Alteration to the dopamine replacement regimen may sometimes help.

#### Sialorrhoea and dysphagia - treatment recommendations

- ✓ Be aware that cholinesterase inhibitors and antipsychotics such as quetiapine and clozapine can aggravate drooling and dysphagia.
- ✓ Speech and language therapists can provide advice on cues/prompts to deliberately swallow when needed.<sup>45</sup> Expiratory muscle strength training may help some.
- ✓ Thickened fluids may help with the prevention of fluid aspiration.<sup>46</sup>
- ✓ Use of sugar free chewing gum or boiled sweets may help some by stimulating swallowing.<sup>47</sup>
- ✓ Anticholinergics, should not generally be used to treat sialorrhoea in LBD.<sup>48</sup>
- ✓ Botulinum toxin injections to the salivary glands are an effective treatment.
- ✓ Clonidine 150 mcg / day is an alternative option but can aggravate orthostatic hypotension and precipitate daytime somnolence.

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<sup>45</sup> An example would be use of a portable metronomic brooch as a reminder to swallow saliva regularly.

<sup>46</sup> Carbonated drinks may also help with swallowing dysfunction.

<sup>47</sup> Be cautious using gum or sweets in patients with swallowing difficulties and/or moderate severe dementia given choking hazard.

<sup>48</sup> Glycopyrrolate is a suggested treatment for sialorrhoea in Parkinson’s disease in the 2017 NICE Guidelines on the management of Parkinson’s disease. However its use in LBD was not supported by the consensus panel.



## Part 8 Managing sleep disturbances

### General principles

Sleep disturbances in LBD include: insomnia, sleep fragmentation, REM sleep behaviour disorder (RBD), motor-related sleep disturbances, restless legs syndrome (RLS), periodic limb movements, obstructive sleep apnoea (OSA), and excessive daytime sleepiness.

The evidence base for assessment and management of sleep disturbances is limited in LBD. However similar sleep symptoms occur in Parkinson's disease and there are a number of general principles for management of sleep disturbances detailed in NICE guidance for the management of Parkinson's Disease (NG71) which have been adapted here for LBD.

### Insomnia and sleep fragmentation

- ✓ A full sleep history should be taken from people with LBD who report sleep disturbance to determine if there is a specific underlying sleep disorder which may be treatable e.g. apnoea, REM sleep behaviour disorder etc.
- ✓ Good sleep hygiene should be advised in people with LBD with any sleep disturbance and includes:
  - avoidance of stimulants (for example, coffee, tea) in the evening.
  - establishment of a regular pattern of sleep, comfortable bedding and temperature.
  - restriction of daytime naps.
  - advice about taking regular and appropriate exercise to induce better sleep.<sup>49</sup>
  - The provision of assistive devices, such as a bed lever or rails to aid with moving and turning, allowing the person to get more comfortable.<sup>50</sup>
- ✓ Review of all medication and avoidance of any drugs that may affect sleep or alertness, or may interact with other medication (for example, cholinesterase inhibitors, selegiline, antihistamines, H2 antagonists, antipsychotics and sedatives).
- ✓ The use of modified/controlled release levodopa at night may be useful if motor symptoms are affecting sleep.
- ✓ Nocturia is a frequent cause of disturbed sleep; however, avoid the use of anticholinergics to treat this (See section Urinary incontinence protocol).<sup>51</sup>
- ✓ Melatonin may improve subjective sleep disturbances in some patients and appears to be well tolerated although there is no evidence to support its use in LBD. Typical dosing is 3mg to 12mg prior to bedtime.
- ✓ Z-drugs<sup>52</sup> may be used short-term for insomnia in extenuating circumstances after ruling out or treating obstructive sleep apnoea; start on lower doses and monitor for potential carry-over side effects of drowsiness, cognitive impairment, falls and confusion.

### REM-Sleep behaviour disorder (RBD)

- ✓ The severity and distress of REM-sleep behaviour disorder can vary between patients; in some circumstances it may not be necessary to treat pharmacologically.<sup>53</sup>

<sup>49</sup> Avoid exercise in the evening, however, as this can potentially activating.

<sup>50</sup> Silk sheets or pyjamas may also help with mobility in bed.

<sup>51</sup> Bottles, commodes or sheath catheters may be options to help manage this.

<sup>52</sup> Often advised as a second-line treatment option.

<sup>53</sup> Some non-dopaminergic medications may worsen RBD e.g. beta-blockers, and consideration should be given to reducing or stopping these.



- ✓ Non-pharmacological strategies for REM-sleep behaviour disorder include lowering bed or placing mattress on the floor, removal of potentially dangerous objects in the bedroom such as sharp or glass objects.
- ✓ It may be necessary for bed partners to sleep separately from the patient.<sup>54</sup>
- ✓ Clonazepam may be helpful. Typical dosing schedules would be 250 mcg – 500 mcg to begin with, with increases up to 1000 mcg, taken 30 minutes prior to bedtime. Caution should be taken in the use of this agent in people with gait disturbance, marked cognitive impairment and/or at significant risk of falls as clonazepam can exacerbate these issues.
- ✓ An alternative to Clonazepam for RBD is melatonin, which has a better side effect profile and is preferred by some clinicians as a first line treatment. Typical dosing is 3mg to 12mg before bedtime.

#### Motor-related sleep disturbances

- ✓ Sleep disturbances arising as result of nocturnal extrapyramidal symptoms (e.g. rigidity, inability to turn in bed etc.) may be improved by the use of long-acting levodopa preparations, for example co-beneldopa or co-careldopa controlled release (CR) 25/100mg taken prior to going to bed.
- ✓ The absorption of long-acting levodopa preparations is variable (between 30-80%) and therefore larger doses therefore may be required to achieve improvement in motor symptoms but this may potentially lead to neuropsychiatric side effects.

#### Restless legs syndrome and periodic limb movements

- ✓ Occasionally restless legs may be secondary to other factors such as iron deficiency anaemia, diabetes or renal dysfunction; it is therefore worth excluding these as causes.
- ✓ In addition, be aware that medications such as antidepressants (esp. serotonergic), antipsychotics and anti-emetics may also exacerbate restless legs syndrome.
- ✓ Regular exercise and avoidance of smoking may help.
- ✓ There is no evidence to support specific treatments of periodic limb movements in LBD. However, similar treatments to restless legs syndrome (see below) can be tried.
- ✓ Dopamine replacement therapy may have a role in the treatment of restless legs syndrome. Ropinirole, pramipexole, and rotigotine have been used to treat moderate to severe restless legs syndrome in LBD.
- ✓ Other agents that have been tried include gabapentin<sup>55</sup> and opioid analgesics; however, the efficacy of these agents in the treatment of restless legs syndrome in LBD is unknown and their use would require a high degree of caution given their potential for cognitive and neuropsychiatric side effects.

#### Obstructive sleep apnoea

- ✓ Assess patients for possible sleep apnoea; ask about pauses in breathing whilst asleep and regular snoring. Excessive daytime somnolence, unrefreshing sleep, early morning headaches and sleep attacks may also be symptoms of sleep apnoea. Risk factors include:
  - Being overweight
  - Male

<sup>54</sup> Consider patient and the bed partner's views on this; on one hand preservation of the relationship may be very important by remaining in the same bed, on the other hand ensuring the bed partner has a restful night's sleep may be more important.

<sup>55</sup> Note gabapentin is a controlled drug in the United Kingdom.

- Smoker
  - Taking medications with sedative effect (e.g. benzodiazepines)
  - Alcohol consumption esp. prior to going to sleep
  - Gastroesophageal reflux
  - Anatomical considerations: narrowed airways, large tongue, tonsils, small jaw or large neck (increased risk in men with collar size > 43cm or 17 inches).
- ✓ If there is a suspicion of sleep apnoea consider referring the patient to a sleep centre for further evaluation and management.

### Excessive daytime sleepiness

- ✓ Document the frequency and occurrence of daytime sleepiness. In some cases the Epworth Sleepiness Scale questionnaire may be helpful in the assessment of this symptom.
- ✓ Give advice on good sleep hygiene, assess for any sleep disturbances and treat as appropriate (see section: Sleep disturbances for further details).
- ✓ Exclude other physical causes e.g. hypothyroidism, anaemia, insomnia, sleep apnoea etc. Be aware that medications commonly used in LBD can cause daytime sleepiness including sedatives, antipsychotics and dopamine replacement therapies - these may need to be reduced or stopped if the excessive daytime sleepiness is severe.
- ✓ Be particular aware of patients who have a sudden onset of sleep – they should be advised not to drive if still doing so.
- ✓ General lifestyle advice should include:
  - Getting adequate exposure to light during the day and darkness at night
  - Engage in physical exercise appropriate to the level of functioning but avoid strenuous activities before bed
  - Social engagement and activities outside the home can provide stimulation to reduce excessive daytime sleepiness.
- ✓ There is limited evidence to support pharmacological interventions for excessive daytime sleepiness in LBD.<sup>56</sup>
- ✓ Improvements in excessive daytime sleepiness may also be seen as a by-product of cholinesterase inhibitor use in some patients.

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<sup>56</sup> Modafinil may be appropriate in selected patients and methylphenidate has been used in PD for excessive daytime sleepiness. However these agents are best prescribed by a clinician experienced in their use.

## Part 9 Acknowledgements

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