Dementia in Dementia with Lewy Bodies May Not Be Attributable to Alzheimer Pathology

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We read with interest the report by Gilman and colleagues concerning the use of [^{11}C]dihydrotetrabenazine positron emission tomography (PET) to differentiate dementia with Lewy bodies (DLB) from Alzheimer's disease (AD).[1] We do, however, have some concerns regarding their use of terminology.

The authors divide DLB into those patients with dementia preceding or starting at the same time as their movement disorder, which they term *DLB-AD*, and those in which parkinsonism preceded the dementia, which they term *DLB-PD*.[1] The term *DLB-AD* implies that the dementia in DLB is attributable or inevitably linked to Alzheimer's disease. Although a degree of Alzheimer pathology often accompanies DLB, genetic studies demonstrate that dementia in Lewy body Parkinson's disease may occur in the absence of Alzheimer pathology,[2] and Alzheimer pathology is not always seen in sporadic DLB: it is notable that in the one case of DLB-AD coming to postmortem in Gilman and colleagues' study, no AD pathology was found.[1] The consensus criteria for the diagnosis of DLB arbitrarily determine that patients in whom dementia is unaccompanied by parkinsonism for the first year be diagnosed with DLB and those in whom motor features occur after the first year as having Parkinson's disease dementia (PDD).[3] It is likely that these diseases are on a continuum, a hypothesis that maybe supported by the similar PET findings in both DLB-AD and DLB-PD reported by Gilman and colleagues.[1] Although the terms *PDD* and *DLB* may be imperfect, we suggest that they may cause less confusion than the terms *DLB-PD* and *DLB-AD*.

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References

1. Gilman S, Koeppe RA, Little R, et al. Striatal monoamine terminals in Lewy body dementia and Alzheimer's disease. Ann Neurol 2004;55:774–780.
2. Zarranz JJ, Alegre J, Gomez-Esteban JC, et al. The new mutation, E46K, of alpha-synuclein causes Parkinson and Lewy body dementia. Ann Neurol 2004;55:164–173.
3. McKeith IG, Galasko D, Kosaka K, et al. Consensus guidelines for the clinical and pathologic diagnosis of dementia with Lewy bodies (DLB): report of the consortium on DLB international workshop. Neurology 1996;47:1113–1124.

DOI: 10.1002/ana.20271

Reply

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Drs Schott, Lees, and Rossor express concern regarding the terminology in our recent report describing the use of [^{11}C]dihydrotetrabenazine with positron emission tomography (PET) to differentiate dementia with Lewy bodies (DLB) from Alzheimer's disease (AD).[1] We divided the DLB cases into two groups, 6 patients who developed parkinsonian features at least 1 year before dementia appeared (DLB/PD) and 14 who developed dementia before parkinsonism or at about the same time (DLB/AD). They consider the abbreviations DLB/AD and DLB/PD to be misleading, in that they regard the term *DLB/AD* as implying that the dementia in DLB "is attributable or inevitably linked to Alzheimer's disease." They would have preferred that we used the consensus guidelines; however, they appear to have misquoted these guidelines.[2] They quote the consensus criteria as recommending the diagnosis of DLB for patients with *dementia without parkinsonism for one year*, and the diagnosis of PDD for patients with *dementia who develop parkinsonism after the first year*. The guidelines state, "… if *dementia occurs within 12 months of the onset of extrapyramidal motor symptoms*, the patient should be assigned a primary diagnosis of possible DLB … If the clinical *history of parkinsonism is longer than 12 months*, PD with dementia … a more appropriate diagnostic label …" (italics are ours).

When we were preparing our report, we discussed at length the terminology recommended by the consensus guidelines and decided against using it even though the definitions of the two groups we described conform to those guidelines. Our reasons were that (1) the definitions of DLB and PDD recommended by the consensus guidelines are purely arbitrary; (2) the terms *DLB* and *PDD* imply different neuropathological underpinnings to the disorders; (3) Alzheimer pathology may or may not accompany widespread Lewy body pathology in both DLB and PDD cases; and (4) apart from comments on advanced AD, the guidelines do not address the time course for presentation of parkinsonian symptoms after initial cognitive decline in DLB. Accordingly, we selected abbreviations that we carefully defined in the article to avoid any implication regarding the neuropathological changes that might be found. As Dr Schott and colleagues found our terminology to be confusing, perhaps we should have used more neutral terms such as DLB-C for patients who develop cognitive disorders in advance of parkinsonian features and DLB-P for those who develop parkinsonian features in advance of dementia.

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References

1. Gilman S, Koeppe RA, Little R, et al. Striatal monoamine terminals in Lewy body dementia and Alzheimer's disease. Ann Neurol 2004;55:774–780.
2. McKeith IG, Galasko D, Kosaka K, et al. Consensus guidelines for the clinical and pathologic diagnosis of dementia with Lewy bodies (DLB): report of the consortium on DLB international workshop. Neurology 1996;47:1113–1124.

10.1002/ana.20273

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