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Letter to the Editor

The incidence and prevalence of dementia with Lewy bodies is underestimated

Vann Jones & O’Brien’s meta-analysis of epidemiological studies (Vann Jones & O’Brien, 2013) reported incidence (0.57–1.4 cases/1000 person-years), and prevalence (3.8–4.5% of all new diagnoses) for dementia with Lewy bodies (DLB). The authors do an excellent job explaining the complexities underlying the significant data variation in the source reports. I would like to expand upon their arguments as to why the true incidence and prevalence may be significantly higher than the reported ranges.

First, as Vann Jones & O’Brien note, under-diagnosis is common: the sensitivity of the clinical diagnosis of DLB is only 32%, whereas specificity is 95% (Nelson et al. 2010). Clinicians in these expert centers suspected DLB, or dual Alzheimer’s/DLB pathology in 8.1% of 2861 cases, yet 14.3% of the cases had significant Lewy pathology at autopsy (Nelson et al. 2010). DLB may be further under-diagnosed outside of specialist dementia settings, where the clinical syndrome is less well-known. Against this we must factor in referral bias. Atypical cases are also more likely to undergo autopsy, and thus enrich the pathology data with atypical findings.

Second, the prevalence increases by 40–300% when possible DLB is included (Vann Jones & O’Brien 2013), and when populations in secondary referral centers are examined.

Third, the ‘1-year rule’ arbitrarily divides these Lewy body diseases: subjects with cognitive impairment within a year of the onset parkinsonism are diagnosed with DLB, whereas those whose cognitive problems begin after 1 year are labeled Parkinson disease dementia (PDD; McKeith et al. 2005; Aarsland et al. 2009). DLB sits at the cusp between movement disorder and behavioral neurology specialty clinics, and referral patterns may dictate diagnosis. Movement disorder specialists underestimate cognitive deficits (Hely et al. 2008) and dementia specialists overlook parkinsonism (Schneider et al. 2007). Yet 42% of incident PD cases have mild cognitive impairment (Yarnall et al. 2013) and 31% of all PD patients are demented (Aarsland et al. 2005), almost triple the rate of controls (de Lau et al. 2005).

Finally, DLB diagnosis can stand or fall on the basis of this core criterion: ‘spontaneous features of parkinsonism’. An unanswered question is this: how much parkinsonism is ‘enough’ parkinsonism? More research is needed.

Declaration of Interest

None.

References


Vann Jones SA, O’Brien JT (2013). The prevalence and incidence of dementia with Lewy bodies: a systematic