Prevalence of dementia in intellectual disability using different diagnostic criteria

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Background Diagnosis of dementia is complex in adults with intellectual disability owing to their pre-existing deficits and different presentation.

Aims To describe the clinical features and prevalence of dementia and its subtypes, and to compare the concurrent validity of dementia criteria in older adults with intellectual disability.

Method The Becoming Older with Learning Disability (BOLD) memory study is a two-stage epidemiological survey of adults with intellectual disability without Down syndrome aged 60 years and older, with comprehensive assessment of people who screen positive. Dementia was diagnosed according to ICD – 10, DSM – IV and DC – LD criteria.

Results The DSM – IV dementia criteria were more inclusive. Diagnosis using ICD – 10 excluded people with even moderate dementia. Clinical subtypes of dementia can be recognised in adults with intellectual disability. Alzheimer’s dementia was the most common, with a prevalence of 8.6% (95% CI 5.2 – 13.0), almost three times greater than expected.

Conclusions Dementia is common in older adults with intellectual disability, but prevalence differs according to the diagnostic criteria used. This has implications for clinical practice.

Declaration of interest None.

METHOD

The Becoming Older with Learning Disability (BOLD) memory study is a two-stage epidemiological survey of dementia in the total population of adults with intellectual disability without Down syndrome aged 60 years and older living in five London Boroughs; this area had a total adult population aged 60 years and older of 177,544 people in the UK 2001 census (http://www.statistics.gov.uk/census2001/census2001.asp). The protocol received approval from the Thames Valley Multi-centre Research Ethics Committee and from the Research and Development offices of all participating National Health Service (NHS) organisations.

Participants We identified potential participants from social services’ electronic databases of past and present intellectual disability service users combined with lists of past or present users of local intellectual disability health teams. We also contacted all residential and day services providers for adults with intellectual disability to ensure that all known older adults with such disability had been identified. Participants included those resident in their own homes, family homes, residential homes of all types, nursing homes and hospitals. In two of the boroughs we also contacted all geriatricians, old age psychiatrists, mental health teams for older people, and all residential and nursing homes caring for people without intellectual disability: this resulted in the identification of only one additional participant with intellectual disability, and so this extension of the sampling frame was not implemented in the other three boroughs. Participants received accessible information written in simple language with pictures. A capacity assessment was undertaken to determine whether the person was able to provide consent; if this was the case, written informed consent was obtained. For those who did not have capacity to consent, assent was given by carers, provided the person did not show unwillingness to participate. Written informed consent was also gained from informants for their own participation. We sought historical information to cover at least the preceding 2 years for those who screened positive.

Intellectual disability was defined according to ICD–10 criteria for mental retardation (World Health Organization,
completed by a qualified intellectual dis-
pleted a full assessment to elicit symptoms
Assessment of people who
schedule based on the Adaptive Behavior
(Strydom & Hassiotis, 2003). They also
psychometric properties in this population
(DMR; Evenhuis, 1996), an established
naire for Persons with Mental Retardation
mants completed the Dementia Question-
of dementia or cognitive decline. Infor-
All participants who were able and all in-
teristic features, and excluded from the
study.
Screening stage
All participants who were able and all in-
forms completed a screen for symptoms of
dementia or cognitive decline. Inform-
ments completed the Dementia Question-
naire for Persons with Mental Retardation
(DMR; Evenhuis, 1996), an established
good screening tool for dementia with good
psychometric properties in this population
(Strydom & Hassiotis, 2003). They also
completed a brief activities of daily living
schedule based on the Adaptive Behavior
Scale (Nihira et al, 1992) and the Activities
for Daily Living Schedule (Lawton &
Brody, 1969). We recorded collected in-
formation about level of functioning in early
life and decline in activities of daily living
over the past 2 years from informants. Par-
ticipants with intellectual disability who
had sufficient communication ability com-
pleted a three-item object memory task
based on the Shoe Box Test (Burr &
Aylward, 2000; Silverman et al, 2004).
Screening criteria were designed for maxi-
mum sensitivity so that no person with
dementia would be missed. Screen positives
fulfilled any of the following conditions: a
score at or above the cognitive score thresh-
olds for dementia provided by Evenhuis
(1996) for severe, high-moderate or mild
intellectual disability on the DMR; an un-
explained decline in activities of daily liv-
ing; or a delayed recall of fewer than two
items in the memory task. Participants
who screened negative on these criteria
were presumed not to have dementia.
Assessment of people who
screened positive
Participants who screened positive com-
pleted a full assessment to elicit symptoms of
dementia as described below.
All screening tests and assessments were
completed by a qualified intellectual dis-
ability psychiatrist (A.S.).
Neuropsychological assessment
Basic neuropsychological assessment con-
sisted of the Test for Severe Impairment
(Albert & Cohen, 1992), additional mem-
ory items from the Severe Impairment Bat-
tery (Saxton & Swihart, 1989), the Tower
of London test (Shallice, 1982), the Super-
market Fluency task (Troyer, 2000), the
British Picture Vocabulary Scale (Dunn &
Dunn, 1997) and the Luria three-stage
command (Hodges, 1994). Informants also
completed a questionnaire based on a mod-
ification of the Cambridge Mental Disor-
ders Examination (CAMDEX) informant
questionnaire to elicit a history of changes
in memory, personality, general cognitive
function and confusion (Ball et al, 2004).
Physical examination
A structured physical examination was con-
ducted to record neurological symptoms
and signs associated with dementia and to
identify other physical disease such as
thyroid disease, neurological conditions
and cardiovascular disorders, based on
memory clinic assessments (Hassiotis et al,
2003). This included a vision and hearing
screen. Informants provided details of cur-
rent health and medications, and medical
records were reviewed to obtain infor-
mation on previous health status and recent
investigations. We recorded the results of
neuroimaging undertaken in the preceding
2 years.
Mental state examination
Mental disorders and psychiatric symptoms
were screened for with the mini Psychiatric
Assessment Schedule for Adults with Devel-
opmental Disability (PAS–ADD), a tool for
assessing adults with intellectual disability
(Moss, 2002).
Diagnosis
All the above information was compiled in
an anonymised summary, which was pre-
sented to two of three psychiatrists (A.H.,
G.L. or A.S.) for independent diagnostic
review. Two were intellectual disability
psychiatrists and one (G.L.) was an old
age psychiatrist. Any disagreement in
ratings was settled by discussion with the
third psychiatrist. A specially developed
tick list with operationalised criteria was
used to produce a differential diagnosis. We
applied the following diagnostic principles:
(a) The key to dementia diagnosis in this
population is decline in cognitive function
from an individual baseline, not change
from a normal level (Aylward et al, 1997).
(b) We followed a hierarchical process,
consistent with diagnostic systems
such as DC-LD (Royal College of
Psychiatrists, 2001), whereby develop-
mental level, mental retardation
syndrome, autistic disorders, physical
disease and medication effects, sensory
loss, environmental change or life
events, or mental illness had to be
considered sequentially as possible
reasons for screening positive.
(c) General dementia criteria had to be met
first before moving on to subtyping.
However, since criteria for Lewy body
dementia and frontotemporal dementia
were designed as stand-alone criteria
outside of the ICD–10 or DSM–IV
criteria, these disorders were not
subjected to the two-stage process.
(d) The list included the following criteria for
dementia: ICD–10 Research Diagnostic
Criteria (World Health Organiza-
tion, 1993), DSM–IV–TR (American
Psychiatric Association, 2000) and
DC-LD criteria (Royal College of
Psychiatrists, 2001), which are
compared in Table 1; ICD–10 (World
Health Organization, 1993), DSM–IV
(American Psychiatric Association,
2000) and the National Institute of
Neurological and Communicative Dis-
orders and Stroke–Association Interna-
tionale pour la Recherche et l’Enseignement
en Neurosciences NINDS–AIREN
(Roman et al, 1993) criteria for
vascular dementia; the Consortium on
Dementia with Lewy Bodies (DLB)
criteria (McKeith et al, 1996); and the
Work Group on Frontotemporal
Dementia and Pick’s Disease criteria
for frontotemporal dementia (FTD;
McKhan et al, 2001).
(e) Dementia is an organic disorder and
should therefore trump mental illnesses
such as depression in hierarchical
systems; instead, it is often defined as
diagnosis of exclusion in the diag-
nostic systems. We made the diagnosis
of dementia in the presence of depres-
sive symptoms if these were deemed
not to account for the cognitive
decline, but the final judgement
We included a clinical rating of memory and other cognitive function, but does not have a separate Impaired domain/symptoms DSM–IV ICD–10 DC–LD

<table>
<thead>
<tr>
<th>Impaired domain/symptoms</th>
<th>DSM–IV</th>
<th>ICD–10</th>
<th>DC–LD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Memory</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Higher cortical functions(^2)</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Executive function</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>Thinking</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>Judgement</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>Other cognitive skills</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Information processing</td>
<td></td>
<td></td>
<td>O</td>
</tr>
<tr>
<td>Aphasia/language skills</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>Apraxia</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>Agnosia</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>Behavioural and emotional function(^2)</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Emotional lability</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>Irritability</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>Apathy</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>Social behaviour</td>
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<td></td>
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<tr>
<td>Other criteria</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Change from premorbid state/decline in level of functioning(^1)</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Duration of at least 6 months</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Exclusions</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Not caused by delirium</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Not caused by mental illness</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
</tbody>
</table>

\(^+\), required for diagnosis.

1. The ICD–10 classification requires a decline in memory and other cognitive function, but does not have a separate criterion for change or deterioration in function.
2. At least one of the circled is required.

For the purpose of this analysis the participants were divided into two groups: those with dementia (if they met any of the above diagnostic criteria) and those who did not meet the criteria.

**Statistical analysis**

Data were entered into the Statistical Package for the Social Sciences version 11 for Windows. Prevalence rates are presented in percentages, rounded to one decimal place. Symmetrical exact binomial 95% confidence intervals were derived using a calculator available at http://statpages.org/confit.html. Chi-squared tests were used to analyse categorical variables with continuity correction for 2 x 2 tables; Fisher’s exact tests were used if 50% or more cells had expected values of less than 5. Significance level was set at \( P < 0.01 \) owing to the number of tests; \( t \)-tests were used to analyse differences in mean age. Correlation between sets of criteria was calculated with Spearman’s rho.

Prevalence rates for Alzheimer’s disease and vascular dementia in the general population were obtained from the most recent European collaborative study of population-based cohorts (4.4% for Alzheimer’s disease and 1.6% for vascular dementia; Lobo et al., 2000). These rates were used to calculate expected counts for this study. The observed count divided by the expected count provided standardised morbidity ratios (SMRs) for comparison of rates between populations (Page et al., 1995). Confidence intervals for SMRs were obtained with a calculator providing exact 95% Poisson confidence intervals (http://home.clara.net/sisa/smr.htm).

**RESULTS**

We identified 258 potential participants from health or social services. An additional 23 (8.2%) were identified through other providers. All 281 potential participants were contacted. Of these, 24 (8.5%) were ineligible for the study because of unrecorded Down syndrome status, being too young, having died recently, not having an intellectual disability, or not residing at the given address. Of the remaining 257 individuals, 35 (13.6%) refused participation, or their carers refused on their behalf; 222 (86.4%) participated. The prevalence of eligible participants in the total population of all adults aged 60 years and older living in these boroughs was 0.15%. Participants did not differ from non-participants in terms of mean age (68.8 v. 67.9 years; \( t = 0.776, P = 0.439 \)) or gender (Pearson \( \chi^2 = 0.14, P = 0.708 \)). The proportion of male to female participants was 52.7% to 47.3%. With regard to severity of disability, 123 (55.4%) participants were rated to have mild intellectual disability and 99 (44.6%) had moderate or more severe disability.

**Participants who screened positive**

Overall, 60 people screened positive; 29 of these met at least one set of dementia criteria (including DLB and FTD criteria). Of these, 13 (45%, or 5.9% of the total) already had the diagnosis of probable or possible dementia recorded in their clinical notes. ‘False’ positives (i.e., those who screened positive but did not meet dementia criteria) were younger (mean age 70.9 v. 76.4 years; \( t = -2.667, P = 0.01 \)) and more likely to have severe intellectual disability (41.9 v. 3.4%; \( \chi^2 = 10.349, P = 0.001 \)), but the true positives and false positives did not differ significantly with...
regard to gender, health problems, mental illness or sensory disabilities.

### Mental illness

The prevalence rates of current mental illness (as reported by informants or extracted from medical records) are given in Table 2; this table also includes the numbers with scores above the mini PAS–ADD thresholds. The proportions of those with mental illness who were also diagnosed with dementia are given in the last column. Since depression is an important differential diagnosis of dementia and may be difficult to distinguish from dementia in older adults, we examined all the cases with a history or mini PAS–ADD threshold score of depression that also met the criteria for dementia. Six adults with a recent history of depression were deemed to have dementia. Only two of them had scores above the depression threshold of the mini PAS–ADD; the rest had fully recovered or had remission of most symptoms, and their cognitive declines were deemed not to relate to the depressive episode. Three of them met all three sets of dementia criteria; one met only the DSM–IV criteria because she did not have a history of behavioural or social decline. She was diagnosed with dementia due to Parkinson’s disease. Of the adults who reached the mini PAS–ADD threshold for depression, one was a 69-year-old woman with mild intellectual disability and a long history of cognitive decline, considerable loss of function and emergence of other neuropsychiatric symptoms. She was diagnosed by her local intellectual disability psychiatrist as having Alzheimer’s disease 2 years prior to participating in the study, and was treated with donepezil for 6 months. She was rated to have depression, symptoms secondary to dementia and met the dementia criteria of ICD–10, DC–LD and DSM–IV. The other person was a 75-year-old man with mild intellectual disability and a history of psychotic illness with depressive episodes since early adulthood. He had a 2-year history of gradual decline in cognitive function and activities of daily living, personality and behavioural changes, episodes of confusion and falls. He had memory deficits on psychometric testing and met the ICD–10 criteria for dementia, but not those of DSM–IV or DC–LD because the raters were not unable to exclude the possibility that his symptoms were related to his mental illness.

### Dementia symptoms

There were 26 participants with dementia for whom the informants could identify the initial symptoms. The most common initial symptom was general deterioration in functioning (n=13; 50% of those with dementia), followed by behavioural or emotional change (n=4; 15.4%). Deterioration in memory (n=2; 7.7%) or other cognitive functions (n=2; 7.7%) was rarely noticed to be prominent in the early stages of the disorder. Other early symptoms (n=5) included episodes of confusion (n=3).

We compared the current dementia symptoms reported by informants in those who screened positive by diagnostic group (any dementia compared with no dementia) (Table 3). The most common reported symptoms for those with dementia were decline in self-care (90% of those with dementia), decline in instrumental activities of daily living (72%), memory decline (73%), episodes of confusion (52%) and the development of muddled thinking (62%). Symptoms that significantly discriminated between those with and without dementia in those who screened positive were deterioration in self-care ability, deterioration in instrumental activities of daily living, change in memory, development of muddled thinking, development of problems with thinking ahead and planning, and newly developed perseveration. None of the behavioural and emotional symptoms was discriminative of dementia.

### Overall dementia and subtype prevalence rates

Prevalence rates for dementia and subtype criteria are given in Table 4. Criteria for Alzheimer’s disease (ICD–10, DSM–IV or NINCDS–ADRDA) were met in 66% of those with dementia. The second most common subtype was Lewy body dementia (possible and probable cases) followed by frontotemporal dementia and then vascular dementia. Frontotemporal dementia was the most common subtype after Alzheimer’s disease if possible cases of Lewy body dementia are discounted. Alzheimer’s and vascular dementias diagnosed by DSM–IV criteria were almost twice as common as the corresponding ICD–10 rates (Table 4). The prevalence rates for those aged 65 years or over who met any criteria for Alzheimer’s or vascular dementia were used to make comparisons with the general population rates. The 17 observed cases of Alzheimer’s disease among those aged 65 years or over compared with 6.25 expected cases resulted in a standardised morbidity ratio (SMR) of 2.72 (95% CI 1.58–4.35). The corresponding observed v. expected count for vascular dementia was 5 v. 2.27 (SMR=2.20, 95% CI 0.72–5.14).

### Dementia criteria

Twenty-eight people met any of the ICD–10, DSM–IV or DC–LD criteria for dementia; 27 of these (12.2% of the total sample)
Table 3  Dementia symptoms reported by informants (screen-positive cases; n=60)

<table>
<thead>
<tr>
<th></th>
<th>No dementia</th>
<th>Dementia on any criteria</th>
<th>Within dementia group %</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n=31</td>
<td>n=29</td>
<td></td>
</tr>
<tr>
<td></td>
<td>n (%)</td>
<td>n (%)</td>
<td></td>
</tr>
<tr>
<td>Memory</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Change in memory**</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>28 (80)</td>
<td>7 (20)</td>
<td>27</td>
</tr>
<tr>
<td>Yes</td>
<td>0 (0)</td>
<td>19 (100)</td>
<td>73</td>
</tr>
<tr>
<td>Executive function</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Decision difficulty</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>24 (56)</td>
<td>19 (44)</td>
<td>76</td>
</tr>
<tr>
<td>Yes</td>
<td>0 (0)</td>
<td>6 (100)</td>
<td>24</td>
</tr>
<tr>
<td>Thinking ahead/planning problems**</td>
<td></td>
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<td></td>
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<tr>
<td>No</td>
<td>26 (63)</td>
<td>15 (37)</td>
<td>54</td>
</tr>
<tr>
<td>Yes</td>
<td>1 (7)</td>
<td>13 (93)</td>
<td>46</td>
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<tr>
<td>Other cognitive functions</td>
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<td></td>
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<tr>
<td>Keep mind on things/concentration</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>24 (62)</td>
<td>15 (38)</td>
<td>54</td>
</tr>
<tr>
<td>Yes</td>
<td>5 (28)</td>
<td>13 (72)</td>
<td>46</td>
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<tr>
<td>Muddled thinking**</td>
<td></td>
<td></td>
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<tr>
<td>No</td>
<td>22 (69)</td>
<td>10 (31)</td>
<td>36</td>
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<tr>
<td>Yes</td>
<td>5 (22)</td>
<td>18 (78)</td>
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<tr>
<td>Talking more or less</td>
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<tr>
<td>No</td>
<td>22 (59)</td>
<td>15 (41)</td>
<td>52</td>
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<tr>
<td>Yes</td>
<td>9 (39)</td>
<td>14 (61)</td>
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<td>Word-finding difficulty</td>
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<tr>
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<td>28 (61)</td>
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<tr>
<td>Yes</td>
<td>1 (10)</td>
<td>9 (90)</td>
<td>33</td>
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<tr>
<td>Perseveration**</td>
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<tr>
<td>No</td>
<td>29 (62)</td>
<td>18 (38)</td>
<td>64</td>
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<td>1 (9)</td>
<td>10 (91)</td>
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<td>Behavioral and emotional functions</td>
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<tr>
<td>More impulsive</td>
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<td>27 (51)</td>
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<td>Other symptoms</td>
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<td>Hallucinations</td>
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<td>3 (33)</td>
<td>6 (67)</td>
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<td>Episodes of confusion</td>
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<td>25 (64)</td>
<td>14 (35)</td>
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</tr>
<tr>
<td>Yes</td>
<td>6 (29)</td>
<td>15 (71)</td>
<td>52</td>
</tr>
</tbody>
</table>

ADL, activities of daily living.

**P < 0.001; z² tests with continuity correction and 1 degree of freedom.

met the criteria for DSM-IV dementia, 22 (9.9%) met the criteria for ICD-10 dementia and 23 (10.4%) the criteria for DC-LD dementia. The overlap between these criteria is shown in Fig. 1: this demonstrates that 21 participants (75%) met all three sets of criteria, those meeting DC-LD criteria were a subset of those meeting DSM-IV criteria, and there were 5 participants who met one set of diagnostic criteria only (ICD-10 or DSM-IV). The criteria are therefore correlated as follows: DSM-IV x ICD-10 r=0.772 (P<0.005); DSM-IV x DC-LD r=0.872 (P<0.005); DC-LD x ICD-10 r=0.894 (P<0.005).

The raters made clinical ratings of severity of dementia for all 29 meeting at least one set of criteria: 12 (41%) were rated as having mild dementia, 16 (55%) as having moderate dementia and 1 (3%) as having severe dementia. Those diagnosed according to ICD-10 and DSM-IV dementia criteria were compared according to severity of dementia. Ten (83%) of the 12 rated as having mild dementia met DSM-IV criteria compared with 8 (66.7%) who met ICD-10 criteria. Six people met criteria for DSM-IV dementia but not ICD-10, and one met the criteria for ICD-10 but not DSM-IV. Of the six diagnosed by DSM-IV but not by ICD-10, half were rated clinically to have dementia of moderate severity. These were
excluded from ICD–10 criteria either because informant history of memory decline was absent (as opposed to other evidence of such decline, which is acceptable for DSM–IV diagnosis) or by the absence of behavioural and emotional symptoms. The extra ICD–10 case was rated to have mild dementia. The reason this did not meet DSM–IV criteria was that depressive symptoms were present and therefore one of the DSM–IV exclusion criteria was met.

**DISCUSSION**

This is the first study to report the prevalence of subtypes of dementia, including frontotemporal and Lewy body dementia, in older adults with intellectual disability. We have demonstrated that the symptoms associated with all dementia subtypes can be recognised in older adults with such disability. As in their general population counterparts, Alzheimer’s disease was the most common diagnosis, but with a prevalence of almost three times higher than expected.

Because dementia may present differently in this population compared with the general population, criteria for the disorder may also perform differently. This is the first study to make a detailed comparison of dementia criteria in older adults with intellectual disability. We have demonstrated that correlations between the ICD–10, DSM–IV and DC–LD dementia criteria were good, but there were important differences. The DSM–IV criteria diagnosed a larger number of participants with mild dementia than ICD–10 criteria and were therefore more inclusive. The ICD–10 criteria excluded not only those with mild dementia, but also a considerable proportion of those with moderate-to-severe dementia.

** Limitations**

This study is the largest cross-sectional survey of dementia in the intellectual disability population to date; our sample represents approximately 1% of the estimated 26 000 adults aged 60 years and over known to have intellectual disability in England (Emerson & Hatton, 2004). We employed epidemiological sampling methods and achieved high participation rates. We identified all older adults known to have intellectual disability. Participants underwent a very sensitive screening strategy, and were fully assessed with established assessment methods and tools if screened positive, before we applied a rigorous diagnostic procedure, which incorporated the main diagnostic criteria for dementia.

Despite the comprehensive recruitment strategy, it is possible that we have missed some older adults with intellectual disability who were unknown to social or health services. However, we believe this number

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**Table 4 Prevalence rates for dementia subtypes**

<table>
<thead>
<tr>
<th>Dementia subtype</th>
<th>Age ≥ 60 years (n=222)</th>
<th>Age ≥ 65 years (n=142)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n</td>
<td>%</td>
</tr>
<tr>
<td>Alzheimer's dementia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Any criteria</td>
<td>19</td>
<td>8.6</td>
</tr>
<tr>
<td>Specific criteria</td>
<td></td>
<td></td>
</tr>
<tr>
<td>ICD–10</td>
<td>8</td>
<td>3.6</td>
</tr>
<tr>
<td>DSM–IV</td>
<td>14</td>
<td>6.3</td>
</tr>
<tr>
<td>NINCDS–ADRDAN</td>
<td>12</td>
<td>5.5</td>
</tr>
<tr>
<td>Vascular dementia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Any criteria</td>
<td>6</td>
<td>2.7</td>
</tr>
<tr>
<td>Specific criteria</td>
<td></td>
<td></td>
</tr>
<tr>
<td>ICD–10</td>
<td>3</td>
<td>1.4</td>
</tr>
<tr>
<td>DSM–IV</td>
<td>6</td>
<td>2.7</td>
</tr>
<tr>
<td>NINDS–AIREN</td>
<td>6</td>
<td>2.7</td>
</tr>
<tr>
<td>Dementia of Lewy body type</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Any criteria</td>
<td>13</td>
<td>5.9</td>
</tr>
<tr>
<td>DBL Consortium criteria</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Possible</td>
<td>9</td>
<td>4.1</td>
</tr>
<tr>
<td>Probable</td>
<td>4</td>
<td>1.8</td>
</tr>
<tr>
<td>Frontotemporal dementia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>FTD Work Group criteria</td>
<td>7</td>
<td>3.2</td>
</tr>
<tr>
<td>Other dementias (e.g. head trauma and Parkinson's disease)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Any criteria</td>
<td>3</td>
<td>1.4</td>
</tr>
<tr>
<td>Any dementia</td>
<td>29</td>
<td>13.1</td>
</tr>
</tbody>
</table>

DLB, dementia with Lewy bodies; FTD, frontotemporal dementia.

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**Fig. 1 Venn diagram of participants diagnosed with dementia on different diagnostic criteria.**

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to be small because older adults with such disability are likely to need assistance owing to the functional problems associated with ageing. This is more likely to be provided by agencies outside the family because informal support networks decrease as people grow older. Furthermore, the comprehensive care system in the UK promotes formal assistance. A small number of potential participants unknown to any service might reduce the increased prevalence of Alzheimer’s disease when compared with the general population, but is unlikely to change our main findings about the relative prevalence of subtypes, presentation of dementia or performance of diagnostic criteria. We excluded adults with Down syndrome recognised by their clinical features, but did not undertake chromosomal analysis; it is therefore possible that some of these excluded adults did not have trisomy 21.

Another limitation is that cross-sectional assessments are less reliable than sequential assessments. We therefore supplemented our data with historical information from informants and medical records. Nevertheless, for a proportion of participants we were unable to decide whether or not they had dementia owing to insufficient data; these were included in the group without dementia. Our study might therefore have underestimated the true prevalence of subtypes of dementia.

Dementia symptoms and concurrent validity of dementia criteria

Because of diagnostic difficulties in this population, clinical diagnosis cannot be used as the gold standard for comparison. We therefore determined the correlation of different dementia criteria and demonstrated their utility, but also highlighted particular issues. Cognitive deficits are difficult to demonstrate in adults with limited verbal and functional ability (Burt & Ayward, 1999); clinicians therefore often rely on informant reports of change. Our data confirm that change in memory and higher functions are not noticed early in people with intellectual disability, and because these changes are required for dementia diagnosis, adults with both intellectual disability and dementia may be diagnosed later in the course of the disorder when these changes have become more apparent.

Dementia criteria differ considerably and therefore yield widely differing prevalence rates in the general population (Ballard & Bannister, 2005). The ICD–10 criteria are more specific but less sensitive than DSM–III–R or DSM–IV criteria (Erkinjuntti et al., 1997). We have shown that this is also the case in older adults with intellectual disability. One of the reasons for this is that ICD–10 criteria are more demanding to apply because they are more dependent on reliable information from informants (Henderson et al., 1994). Another limitation of the ICD–10 and DC–LD criteria is that behavioural and emotional changes are an additional required symptom for ICD–10 and DC–LD dementia, but not for DSM–IV dementia. These were reported to have occurred early in a small but significant number of adults with intellectual disability and dementia. However, these symptoms were not good at discriminating between those with and without dementia, and limit the number of people diagnosed with ICD–10 criteria. Even those clinically rated to have moderate severity of dementia did not meet ICD–10 criteria. This was contrary to the expectation of an international consensus group (Ayward et al., 1997).

The ‘false’ screen positives need special mention. Those with severe intellectual disability were more likely to meet screening criteria but not diagnostic criteria for dementia. The proportion of false screen positives may seem high, but a recent study in an elderly population noted that of 96 people with confirmed cognitive and functional impairment, only 55 satisfied the DSM–IV criteria for dementia (Shaji et al., 2005). These authors felt that the DSM–IV prevalence of dementia is possibly an underestimation; this might also be the case in the population with intellectual disability, because the ‘false’ screen positive group might contain cases of dementia that did not meet criteria owing to lack of informant or medical history, or to the difficulty of making this diagnosis in a group with severe disability.

Clinical implications

We found that more than double the number of older adults with intellectual disability meet dementia criteria than is recognised by their carers or health professionals. Functional decline was reported to be more common than memory decline early on in the presentation; perhaps the potential for pathological causes underlying such decline is not recognised in adults with lifelong deficits. Dementia should always be considered as a possible diagnosis when investigating reports of decline in older adults with intellectual disability. Our findings also give credence to screening approaches that rely on functional change (Prasher et al., 2004).

We preferred the DSM–IV criteria for dementia in this population. They are clearly set out and easy to interpret. They do not rely exclusively on informant report of memory and cognitive change like the ICD–10 criteria, which allows the clinician to use other sources of information such as sequential cognitive assessments and medical records. Furthermore, they do not require behavioural or emotional change but focus on functional change, which is important in this population. This has important implications for patients, since the use of DSM–IV criteria may enable earlier diagnosis of dementia in larger numbers of older adults with intellectual disability, which could gain them timely access to appropriate interventions.

Future research

Our findings raise questions about the aetiology of dementia in older adults with intellectual disability but without Down syndrome. It is important to establish why Alzheimer’s dementia may be more common in these adults than in the general population; we have estimated an SMR of 2.72 (95% CI 1.58–4.35). Possibilities include genetic causes such as apolipoprotein E4 alleles, or environmental causes such as brain damage during birth and early life, which is associated with intellectual disability but may also in the long term be associated with Alzheimer’s disease.

The incidence and presentation of dementia and validity of diagnoses should be confirmed longitudinally. It is also important to confirm subtype diagnoses with post-mortem studies, and to investigate the aetiology of dementia in this population. This will enable appropriate interventions and illuminate our understanding of dementia presentation and progression throughout the intellectual range.

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