The prevalence and causes of dementia in people under the age of 65 years

R J Harvey, M Skelton-Robinson, M N Rossor

Methods

Ethical approval was obtained from the five local research ethics committees covering the catchment areas of the London Borough of Hillingdon (110 km², population 232 000, of whom 81 184 were between 30 and 64 years of age; Jarman Index 21) and the London boroughs of Kensington, Chelsea, and Westminster (25 km², population 355 500 of whom 112 309 were between 30 and 64 years of age; Jarman Index 21).

Sampling

Stage 1

The aim of the first stage of sampling was to raise awareness of the project, and the need to identify and report any case, or suspected case of dementia, where the illness had started before the age of 65 years.

Personal letters and information were sent out to every general practitioner, psychiatrist, old age psychiatrist, neurologist, geriatrician, and general physician. In addition, information was also provided to departments of social services, voluntary organisations, day centres, home care, and community care teams through senior managers in the relevant organisations. The project ran in parallel with social services/voluntary sector initiatives in both areas, providing senior organisational leadership across health and social services and ensuring widespread engagement with the project. Clinicians and practitioners were asked to notify any cases under their care on a defined “census” day for the study.

In parallel, hospital information systems and case registers at the four main hospitals in the catchment areas were searched for cases falling in the target age range who had relevant ICD-9/10 diagnoses.

Personal contact was made with key neurologists and psychiatrists working in the area who allowed hand searching of copies of clinic letters, discharge summaries, and departmental databases.

The study confirms previous “guestimates” of the number of younger people affected by dementia in UK. The prevalence figures generated are robust, and are supported by other smaller and targeted prevalence surveys. The prevalence figures provided by this study will allow health planners to accurately estimate need and plan services.

In the past decade, interest and concern for younger people with dementia have dramatically increased. Following sustained campaigning and support from the Alzheimer’s Society and other voluntary organisations in the UK, the needs of younger people with dementia and those who care for them have been increasingly addressed. The Royal College of Psychiatrists has recently issued guidelines recommending that every area in the country has a named old age psychiatrist responsible for younger people with dementia. This has been matched by the development of specialist support services, day care, and care homes for this group of patients.

However, delivering effective services relies on accurate epidemiological data to drive the planning and implementation process. In the past, estimates of the prevalence of dementia in younger people have relied on historical data drawn from hospital inpatient data and diagnostic registers. The inherent problem of studies based upon known, diagnosed cases is the potential for underestimating the real prevalence, particularly given the rarity and relatively small numbers involved. Unfortunately, because of the relatively low prevalence, conventional “door knocking” would be too costly and time consuming.

The aim of our study was to use a large, geographical catchment area and to use a combination of techniques to identify every known case of dementia starting before the age of 65 years. Having identified cases, we were also interested in exploring the prevalence of the different causes of dementia, and using the results obtained to extrapolate estimates of the numbers of younger people with dementia in the UK.
Stage 2
An experienced psychiatrist (RJH) followed up notification of cases, requesting copies and/or access to all available hospital and GP case notes. The available information included clinical history, and investigations (including neuroimaging) were reviewed.

A hierarchical diagnostic algorithm was then applied. At the top level, subjects achieved a definite diagnosis if the dementia was associated with a known disease-specific genetic mutation, if there were neuropathological results available from a cerebral biopsy, or from autopsy if the patient had died after the “census” date.

At the second level of the algorithm, well recognised and validated clinical diagnostic criteria were applied to make the diagnoses of Alzheimer’s disease (NINCDS/ADRDA criteria), vascular dementia (NINDS/AIREN criteria), dementia with Lewy bodies, frontotemporal dementia (Mand and Chesters Criteria), alcohol related dementia (DSM-IV criteria for alcohol induced persisting dementia), or other dementias for which DSM-IV criteria are available. These criteria were not applied hierarchically, but rather on the basis of best clinical opinion.

At the third level of the algorithm, patients fulfilling the DSM-IV criteria for dementia, but not fulfilling criteria for one of the above diagnostic categories were assigned to a Dementia Not Otherwise Specified category.

The age at onset of disease was defined as the age of the patient at which the earliest conclusive dementia symptom was noted or documented in the medical notes or other correspondence.

Stage 3
For half of the patients identified in stage 2, a direct approach was made through the general practitioner seeking consent for a detailed clinical assessment to confirm diagnosis. Those patients and carers willing to participate were visited at home for a detailed clinical assessment to confirm diagnosis. Those whom the general practitioner could not be contacted or who refused to participate were approached directly by RJH, who completed a semi-structured interview to collect additional information.

For half of the patients identified in stage 2, a direct approach was made through the general practitioner seeking consent and of these, those who responded or who refused to participate were visited at home for a detailed clinical assessment to confirm diagnosis. Those whom the general practitioner could not be contacted or who refused to participate were approached directly by RJH, who completed a semi-structured interview to collect additional information.

Any diagnoses that were not clear at either stage 2 or stage 3 were discussed and agreed by consensus between RJH and MNR.

RESULTS
In total, 227 people with suspected young onset dementia were identified to the study. The sources of identification were psychologists (21%), databases and case registers (20%), neurologists (17%), psychiatrists (14%), social workers (9%), GPs (6%), physicians (6%), community nurses (4%), and other (3%).

Of the 227 referrals, 19 were excluded as not fulfilling DSM-IV criteria for dementia (six were “worried well”, seven had traumatic brain injuries, two had chronic schizophrenia, and four cases identified from hospital information systems were clearly miscoded when the notes were reviewed).

Of the remaining 208 cases, review of the notes showed an age of onset greater than 65 years of age in 23 cases who were further excluded from the study. The remaining 185 cases formed the population for the prevalence study, and of these, 87 cases (47%) underwent a detailed stage 3 personal assessment.

In the stage 3 assessment group, 69 patients (83%) had undergone neuroimaging with either CT or MRI, with 18 (21%) having undergone both CT and MRI. Twenty one patients (24%) had had an EEG.

The mean age of the population was 58.7 years (95% CI 57.4 to 60.1 years). Fifty eight percent (95% CI 50.9 to 65.6%) were male and 42% (95% CI 34.4 to 49.1%) were female, suggesting a trend towards a higher prevalence of dementia in males than females.

Table 1 summarises the age and gender specific prevalence rates for dementia in the study population on the “census” day; the pattern of prevalence approximately doubles with each five year increase in age.

Pragmatically, mixed diagnoses were avoided and we aimed to make a “highest confidence” single diagnosis in each case. In 168 (90%) of cases a single diagnosis was established using the diagnostic algorithm, 19 cases (10%) fitted more than one set of criteria and required a clinical consensus diagnosis to be made. Differential diagnoses in the population included Alzheimer’s disease (62 cases, 34%), vascular dementia (34 cases, 18%), frontotemporal dementia (23 cases, 12%), alcohol related dementia (19 cases, 10%), and dementia with Lewy bodies (12 cases, 7%). Other causes of dementia accounted for 35 cases (19%) and included Huntington’s disease (nine cases, 5%), or other dementias for which DSM-IV criteria are available. These criteria were not applied hierarchically, but rather on the basis of best clinical opinion.

At the third level of the algorithm, patients fulfilling the DSM-IV criteria for dementia, but not fulfilling criteria for one of the above diagnostic categories were assigned to a Dementia Not Otherwise Specified category.

Table 1: Age and gender specific prevalence rates in the study population

<table>
<thead>
<tr>
<th>Population</th>
<th>All causes of dementia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age range</td>
<td>Male (n)</td>
</tr>
<tr>
<td>30–34</td>
<td>23898</td>
</tr>
<tr>
<td>35–39</td>
<td>18526</td>
</tr>
<tr>
<td>40–44</td>
<td>18982</td>
</tr>
<tr>
<td>45–49</td>
<td>16549</td>
</tr>
<tr>
<td>50–54</td>
<td>15185</td>
</tr>
<tr>
<td>55–59</td>
<td>13983</td>
</tr>
<tr>
<td>60–64</td>
<td>12716</td>
</tr>
<tr>
<td>Over 65‡</td>
<td>55</td>
</tr>
</tbody>
</table>

*Rate per 100 000 people at risk.
†Significance of difference between genders by inference from 95% CI.
‡Subjects who had a dementia starting before the age of 65 years, but were over 65 but still living on the study census day.

All Male Female
cases), dementia in multiple sclerosis (eight cases), cortico-
basal degeneration (two cases), prion disease (two cases),
dementia in Down’s syndrome (three cases), dementia in Par-
kinson’s disease (two cases), and dementia not otherwise
specified (nine cases). The very low prevalence of dementia in
Down’s syndrome is likely to be an artefact due to sampling
bias. Despite raising awareness of the project with learning
disability services, no referrals of cases were made from these
sources.

The age specific prevalence rates for the four most common
diseases in this group of younger people are
summarised in table 2.

### Estimates of numbers of cases in the UK

One of the objectives of the study was to generate robust esti-
mates of the numbers of people in the UK with young onset
dementia. These figures are important both for service
planning and for campaigning for improvements in recogni-
tion and provision.

Using data from the 2001 UK Census, we applied the preva-
ience rates derived from our population to the population
figures for the UK. Table 3 summarises the estimated numbers
of younger people with dementia, with specific diagnoses
in the UK.

### DISCUSSION

This is the first study to report the prevalence of dementia and
diagnostic differences in a community based catchment area
study of younger people with dementia. The prevalence of
dementia in people between the ages of 30 and 64 years was 54
per 100 000 with the prevalence approximately doubling for
each five year age band. Based on the prevalence figures
derived from this study we estimate that there are 18 319
(15 296–21 758) people under the age of 65 years with
dementia in the UK.

The main strength of the study is the combination of a large
catchment area and the use of multiple and varied means of
case identification, essentially attempting to leave no stone
unturned. The very varied sources of case identification
confirm the benefits of this methodology. The main weakness
of the study is that despite using a catchment area with a
population of more than 500 000 people, the rarity of the con-
dition meant that only 185 cases were identified. For more
robust estimates with smaller confidence intervals a much
larger population would be helpful.

This study was unique in being large enough to consider a
range of differential diagnoses with our disease specific figures
and overall figures being supported by other studies of
individual diseases.

Thus our prevalence figure for young onset Alzheimer’s dis-
ease of 35.0 per 100 000 in the 45–64 years age group is very
similar to estimates for the Northern Region (34.6),
Finland (32.7), the Framingham Study (31.8),
and Cambridge (15.1).

Similarly, the prevalence rates we found for vascular
dementia were close to the estimates derived from an earlier
study in Scotland, and the prevalence rate we identified for
frontotemporal dementia (15.4 per 100 000) was almost iden-
tical to the rate identified in the Cambridge study (15.1).

While none of these studies used an identical methodology,
this study is probably most similar to the study from the
Northern Region which used case ascertainment from hos-
pital inpatient records, and the Cambridge study which
identified cases from memory clinic databases and admission
records. The Framingham study and the Finnish study both
used ascertainment from large population diagnostic regis-
ters, but generate similar prevalence figures. One conclusion
from this is that younger people with dementia inevitably
come to medical attention and receive a diagnosis—what is
less certain is what follow up and support these patients are
given after the diagnosis.

Our study underpins and supports initiatives from the
Alzheimer’s Society and the Royal College of Psychiatrists to
secure and improve services for this group of patients. Even
though dementia in younger people is uncommon when com-
pared with dementia in older people, every case is a personal

### Table 2: Age specific prevalence rates for the most common causes of young onset dementia

<table>
<thead>
<tr>
<th>Age range</th>
<th>Alzheimer’s disease</th>
<th>Vascular dementia</th>
<th>Frontotemporal dementia</th>
<th>Alcohol related dementia</th>
</tr>
</thead>
<tbody>
<tr>
<td>30–34</td>
<td>15995210</td>
<td>4894 (3513 to 6634)</td>
<td>2507 (1553 to 3835)</td>
<td>2255 (1290 to 3654)</td>
</tr>
<tr>
<td>35–39</td>
<td>27266185</td>
<td>18319 (15296 to 21758)</td>
<td>2536 (1502 to 4008)</td>
<td>2255 (1290 to 3654)</td>
</tr>
<tr>
<td>40–44</td>
<td>3328210</td>
<td>13728 (1134 to 16514)</td>
<td>2149 (1274 to 3401)</td>
<td>1910 (1092 to 3107)</td>
</tr>
<tr>
<td>45–49</td>
<td>4085090</td>
<td>1208 (729 to 1977)</td>
<td>2174 (1188 to 3862)</td>
<td>1973 (1124 to 3323)</td>
</tr>
</tbody>
</table>

### Table 3: Estimated numbers of cases of dementia in people under the age of 65 in the UK

<table>
<thead>
<tr>
<th>Age</th>
<th>UK Population 2001</th>
<th>All cases of dementia</th>
<th>Alzheimer’s disease</th>
<th>Vascular dementia</th>
<th>Frontotemporal dementia</th>
<th>Alcohol related dementia</th>
</tr>
</thead>
<tbody>
<tr>
<td>30–34</td>
<td>4493585</td>
<td>570 (209 to 1240)</td>
<td>107 (27 to 598)</td>
<td>107 (27 to 598)</td>
<td>107 (27 to 598)</td>
<td>107 (27 to 598)</td>
</tr>
<tr>
<td>35–39</td>
<td>4651306</td>
<td>369 (76 to 1078)</td>
<td>664 (216 to 1552)</td>
<td>133 (34 to 739)</td>
<td>133 (34 to 739)</td>
<td>133 (34 to 739)</td>
</tr>
<tr>
<td>40–44</td>
<td>4151590</td>
<td>645 (257 to 1403)</td>
<td>169 (925 to 2641)</td>
<td>847 (341 to 1743)</td>
<td>847 (341 to 1743)</td>
<td>847 (341 to 1743)</td>
</tr>
<tr>
<td>45–49</td>
<td>3775964</td>
<td>1232 (616 to 2204)</td>
<td>224 (27 to 811)</td>
<td>224 (27 to 811)</td>
<td>224 (27 to 811)</td>
<td>224 (27 to 811)</td>
</tr>
<tr>
<td>50–54</td>
<td>4004037</td>
<td>2523 (1519 to 3939)</td>
<td>664 (216 to 1552)</td>
<td>797 (293 to 1733)</td>
<td>797 (293 to 1733)</td>
<td>797 (293 to 1733)</td>
</tr>
<tr>
<td>55–59</td>
<td>3338861</td>
<td>5079 (3673 to 6878)</td>
<td>169 (925 to 2641)</td>
<td>605 (196 to 1412)</td>
<td>605 (196 to 1412)</td>
<td>605 (196 to 1412)</td>
</tr>
<tr>
<td>60–64</td>
<td>2879948</td>
<td>4789 (3456 to 6451)</td>
<td>2228 (1359 to 3427)</td>
<td>334 (69 to 976)</td>
<td>334 (69 to 976)</td>
<td>334 (69 to 976)</td>
</tr>
<tr>
<td>65–69</td>
<td>27266185</td>
<td>18319 (15296 to 21758)</td>
<td>5918 (4254 to 7989)</td>
<td>2959 (1832 to 4526)</td>
<td>2959 (1832 to 4526)</td>
<td>2959 (1832 to 4526)</td>
</tr>
<tr>
<td>70–74</td>
<td>13995210</td>
<td>13728 (1134 to 16514)</td>
<td>4894 (3513 to 6634)</td>
<td>2507 (1553 to 3835)</td>
<td>2507 (1553 to 3835)</td>
<td>2507 (1553 to 3835)</td>
</tr>
</tbody>
</table>

*95% confidence interval of the estimate.
tragedy for the individual and their family, which often includes young children. The study also highlights the differences between dementia in younger people and dementia in older people. Alzheimer’s disease is considerably less common in this group, accounting for only 34% of cases, whereas in the elderly it would be expected to account for up to 80% of cases. Moreover, unusual diseases such as frontotemporal dementia accounts for 12% of cases, although there is increasing evidence that it is a commonly missed diagnosis in older people. The prevalence data generated by this study can be applied to local population figures to derive reliable estimates of the potential numbers of cases in any area. This should support better planning and delivery of services.

ACKNOWLEDGEMENTS

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Competing interests: none declared

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REFERENCES
3 Quinn, C. The Care Must Be There. London: Dementia Relief Trust, 1996.