

Symptoms of dementia among adults with Down's syndrome: a qualitative study

S. Deb,¹ M. Hare² & L. Prior³

¹ University of Birmingham, Birmingham, UK

² Cardiff University, Cardiff, UK

³ Queen's University Belfast, Belfast, UK

Abstract

Background Dementia is common among adults with Down's syndrome (DS); yet the diagnosis of dementia, particularly in its early stage, can be difficult in this population. One possible reason for this may be the different clinical manifestation of dementia among people with intellectual disabilities.

Aims The aim of this study was to map out the carers' perspective of symptoms of dementia among adults with DS in order to inform the development of an informant-rated screening questionnaire.

Method Unconstrained information from carers of people with DS and dementia regarding the symptoms, particularly the early symptoms of dementia, was gathered using a qualitative methodology. Carers of 24 adults with DS and dementia were interviewed. The interviews were recorded and fully transcribed. The transcripts were then analysed using qualitative software.

Results There appeared to be many similarities in the clinical presentation of dementia in adults with

DS and the non-intellectually disabled general population. Like in the non-intellectually disabled general population, forgetfulness especially, impairment of recent memory combined with a relatively intact distant memory and confusion were common, and presented early in dementia among adults with DS. However, many 'frontal lobe'-related symptoms that are usually manifested later in the process of dementia among the general population were common at an early stage of dementia among adults with DS. A general slowness including slowness in activities and speech, other language problems, loss of interest in activities, social withdrawal, balance problems, sleep problems, loss of pre-existing skills along with the emergence of emotional and behaviour problems were common among adults with DS in our study.

Conclusions This study highlights the similarities in the clinical presentation of dementia among the general population and people with DS with a particular emphasis on the earlier appearance of symptoms associated with the frontal lobe dysfunction among adults with DS.

Keywords carer's perspective, dementia, Down's syndrome, qualitative study, screening instrument, symptoms

Correspondence: Professor Shoumitro Deb, MBBS, FRCPSych, MD, Clinical Professor of Neuropsychiatry & Intellectual Disability, Division of Neuroscience, University of Birmingham, Queen Elizabeth Psychiatric Hospital, Mindelsohn Way, Birmingham B15 2QZ, UK (e-mail: s.deb@bham.ac.uk).

Introduction

Autopsy studies have shown brain changes (neuropathology) similar to that of Alzheimer's disease in almost all adults with Down's syndrome (DS) over the age of 40 years (Mann 1988), a fact that is supported by neuroimaging findings (Deb *et al.* 1992). However, it is estimated that clinically, dementia can be diagnosed in only some 9% of adults with DS in their fourth decade, and around 55% in their sixth decade (Prasher 1995a).

The diagnosis of dementia in people with DS remains difficult, particularly in the early stage of the disease. Cognitive abilities in most people with DS are at a below-average level even before they develop dementia. Therefore, the screening instruments commonly used in the general population such as the Mini Mental State Examination (MMSE) (Folstein *et al.* 1975) cannot be used in this population because of possible floor effect. For similar reasons, direct neuropsychological tests cannot be administered in any meaningful way to most people with DS (Deb & Braganza 1999). Only experienced professionals can administer neuropsychological tests, not the carers (Aylward *et al.* 1997). Given the current health policy emphasis on patient and carer participation in all matters affecting health (Department of Health 2006), this is a somewhat unsatisfactory state of affairs, and serves to underline the need for carer involvement in the early stage of diagnosis of dementia among adults with DS.

Prasher (1995a) described mental deterioration, slowing, confusion, reduced output of speech, and deterioration in gait and personality change as the common early symptoms of dementia among adults with DS. Evenhuis (1990) on the other hand described apathy and withdrawal symptoms as the most common early features of dementia among subjects with both moderate and severe intellectual disabilities (ID). The late features of dementia among adults with ID include severe intellectual deterioration, marked change in personality and mood, loss of sphincter control, onset of epileptic seizures, loss of mobility with increased muscle tone and eventual complete loss of all self-help skills (Prasher 1995b).

Many studies have highlighted the similarities in clinical presentation of Alzheimer's dementia in the

general population and in people with DS using both cross-sectional design (Thase *et al.* 1984; Roeden & Zitman 1995) and prospective design (Lai & Williams 1989; Brugge *et al.* 1994). Holland *et al.* (2000) and Ball *et al.* (2006) highlighted the importance of carer reporting of symptoms of dementia among adults with DS. In a 18-month follow-up study, Holland *et al.* (2000) found that in the initial assessment carers reported changes predominantly in behaviour and personality among adults with DS who later developed dementia. The frontal-like dementia was more prevalent particularly among the younger groups. On the basis of their findings, the authors hypothesized that the frontal lobe functions are first to be compromised with the progressive development of Alzheimer-like neuropathology in people with DS. In another 4-year follow-up study, Oliver *et al.* (1998) found that those adults with DS who developed dementia at the follow-up showed evidence of impaired orientation and visuospatial memory at the beginning of the study, but language function and praxis deteriorated later.

Some of the early symptoms of dementia may be subtle or may present as an exacerbation of the existing behavioural traits or manifest differently in people with DS than they do in non-intellectually disabled people. Only carers will notice these early and sometimes unusual changes in the person's behaviour and only by asking carers can we ensure that such symptoms are included in any case detection instrument. The aim of our study was to develop a questionnaire for screening dementia among those adults with DS who have already developed symptoms of dementia.

Method

We developed a dementia screening questionnaire for adults with DS using a two-stage methodology. In the first stage, we collated the items for the questionnaire by using a focus group and qualitative interviews with 24 carers of people with DS and dementia. In the second stage, we field-tested the newly developed questionnaire and used standard statistical measures to assess its validity and inter-rater, and test-retest reliability. Data relating to the psychometric properties of this questionnaire are

reported in a separate paper (Deb *et al.* 2007). The qualitative methods that were used to collect and to analyse the data are similar to those described in the relevant sections of Hesse-Biber & Leavy's (2006) work. In this paper, we present data from the first qualitative stage of the study.

The qualitative stage of the investigation was conducted in the following stages; (1) focus group discussion to clarify terms of reference; (2) open-ended interviews with carers of people with DS and dementia; and (3) data analysis of the transcribed interviews using established methods of coding.

Focus group

An initial focus group discussion took place among the researchers (S.D. & M.H.), a carer of a person with DS and dementia, and a carer of a person who did not have DS or ID but developed dementia. The focus group discussion served to identify key areas of concern for the subsequent qualitative interviews with carers, and clarified terms of references for the main project. We recorded this discussion in a Sony mini disc audio recorder and listened to the recording several times. This was done in order to gather information regarding different clinical manifestation of dementia in a person with and without ID.

Qualitative interviews and analysis

The qualitative interviews served to gather information on the individual family carer's perception and assessment of behavioural changes in an adult with DS since the onset of dementia. We recruited the patients and their family carers primarily from S.D.'s own outpatient lists of current and previous patients in Cardiff. Further, we used S.D.'s local contacts to identify patients with DS with a confirmed diagnosis of dementia. Additional patients suitable for inclusion in the study were identified from a list of such patients used during a previous study by Deb & Braganza (1999).

The inclusion criteria were: (1) patients with DS; (2) patients with a confirmed clinical diagnosis of dementia according to the modified ICD-10 criteria (Aylward *et al.* 1997; the ICD-10 diagnosis was made based on information gathered from various

sources that included information from the carers, direct examination of the patients and the results of the relevant investigations); and (3) patients with an identified family carer who had known the person for long enough to be able to give information on the development of the dementing process.

We obtained approvals from the Welsh Multi-Centre Research Ethics Committee, the appropriate Local Research Ethics Committees and the Research & Development departments before the study commenced. In all cases, we first approached a key worker to establish if there was a family carer, and if so, the key worker contacted the family on our behalf to introduce the study and to seek consent for participation. We did not contact any family carer directly, in accordance with guidance given by the ethics committee.

We approached a total of 42 family carers for the study. A number of these were found to be unsuitable for several reasons, for example: (1) the patient did not have a confirmed diagnosis of dementia according to the ICD-10 criteria; (2) the patient had moved out of the area since last contact; (3) the patient had died more than 12 months ago; (4) there was no family carer who had known the person for long enough to give information on the early symptoms of dementia; and (5) a family carer of a recently deceased patient did not feel ready to talk about the patient's decline.

Family carers completed and signed a written consent form before taking part in the study. Where possible we received an informal verbal agreement from the person with DS for their family carer to participate in the study. We distributed an information sheet explaining the study to all carers before they were asked to sign the consent form. The sample included family carers of 10 women and 14 men with DS. The qualitative data were analysed simultaneously as the interviews were transcribed and any new data gathered from any new interviews were compared with the already collected data from the previous interviews. This reiterative process allowed us to stop after a certain number of interviews. As with much qualitative research, 24 interviews proved sufficient for analysis by which time the data set became saturated and strong themes had emerged (Hesse-Biber & Leavy 2006). We therefore decided to stop interviewing carers at this stage.

M.H. carried out all the interviews. She held the interviews at places convenient to the carer and each interview took approximately 45 min. Semi-structured interviews that focused on narratives of personal experiences of the carers were conducted. Interviewees were asked to describe their perception of symptoms and behavioural changes associated with the onset of dementia among people with DS whom they cared for. Open-ended questions were designed to collect as much information as possible on the behavioural changes. With the permission of the carer, M.H. recorded the interviews on a Sony audio mini-disk (only one carer refused for the interview to be recorded). Particular emphasis was put on new behaviours and changes to existing behaviours, so that a distinction could be made between pre-existing deficits and new symptoms attributable to the developing dementia.

Most recorded interviews were fully transcribed by the researcher (and some were transcribed by a medical secretary). Initially, a simple concordance programme was used to identify a list of the most frequent terms used in the interviews. Following that, the full interview transcripts were read over repeatedly in combination with listening to the recorded interviews. The coding process was akin to that described by Strauss & Corbin (1990), and was later facilitated by the use of NUDI*ST (N5), a qualitative software data analysis package (Richards 1999). The key categories were then used as nodes for use. The themes and topics derived from this analysis are described in the Results section. Open coding was used to highlight those symptoms mentioned in each interview, and we then examined the frequency with which these symptoms were mentioned throughout the interviews. Manual coding was undertaken and a wall chart was used for visual identification of key symptoms, in particular first symptoms (see Table 1) (Hesse-Biber & Leavy 2006). On the basis of the qualitative data and its analysis, S.D., M.R. and L.P. had repeated discussions about the items gathered from the analysis of the qualitative data in order to select the most clinically relevant items and exclude the repeated items. These are presented in the Results section. It was not, however, possible to differentiate various symptoms of dementia with different weightings placed on them by the carers. We have therefore reported

in Table 1 the relative frequency at which different carers have described different symptoms.

Results

The age of the 24 adults with DS included in this study ranged from 48 to 72 years. One person was aged 48 years, 18 were aged between 50 and 60 years, one was aged 61 years and another 62 years, two were aged 71 years and one was 72 years old. Four people had mild, 16 moderate and four severe ID according to the ICD-10 criteria (WHO 1992).

In Table 2 we have provided some verbatim examples of carers' description of early symptoms of dementia among adults with DS. In Table 3 we have provided differing description of early dementia symptoms in relation to different degrees of ID and gender. We describe below some examples of symptoms as described by the carers, though we have categorized them under clinically meaningful headings.

Forgetfulness/memory problems

Memory problems were the most prominent symptoms particularly in the early stage of the disease. Early symptoms included loss of recent memory (loss of information gathered over the last few days) but relatively intact distant memory (memory of events/information gathered several months/years ago). There were many examples. One person used to phone her mother every Sunday at 6 PM, but at the time of the interview she rarely remembered to do so. The same person used to phone his general practitioner's surgery asking for a repeat prescription, but he could no longer do that although his distant memory was good. He remembered a lot about his family and where he lived, and also remembered all about the 'Beatles'. Some other examples included references to the fact that the person with DS: (1) forgets to take his tablets; (2) forgets to turn up for an appointment (an example of prospective memory impairment); (3) turns on the gas fire but forgets to switch on the ignition (an example of procedural memory impairment); (4) has forgotten the names of familiar staff members (an example of semantic memory impairment);

Table 1 Summary of frequency of carers' report of behaviours among adults with Down's syndrome and dementia

Symptoms	101	102	104	105	106	107	108	109	110	114	115	116	117	118	120	122	123	125	128	
Personal hygiene/dressing																				
Unable to wash/bathe unaided		✓	✓	✓	✓		✓		✓	✓	✓			✓	✓	✓	✓	✓	✓	✓
Fear of water/washing hair										✓	✓									✓
Full nursing care required					✓				✓	✓	✓			✓	✓	✓	✓			
Refusal to change clothes/bathe						✓				✓	✓									
Unable to dress unaided		✓	✓	✓	✓		✓		✓	✓	✓		✓	✓	✓	✓	✓	✓		
Inappropriate dressing (back to front, etc.)	✓	✓		✓			☑	✓	✓		✓	✓	✓			✓	✓	☑	✓	✓
Stripping off/exposing							✓			✓						✓	☑			
Eating/drinking																				
Refusing to eat			✓													✓				✓
Overeating/binge eating	✓																			
Unable to use utensils/ needing help		✓	✓		✓				✓	✓	✓		✓	✓	✓	✓	✓	✓		
Unable to swallow/chew										✓					✓	✓	✓	✓		
Needing to be fed			✓		✓				✓	✓	✓		✓	✓	✓	✓	✓	✓		
Toileting																				
Unable to use toilet unaided			✓		✓				✓	✓	✓			✓	✓	✓	✓	✓		
Inappropriate urination (room/corridor)			✓					✓	✓							✓				✓
Soiling/smearing									✓					✓		✓				
Incontinent of urine (incl. accidents)			✓		✓		✓	✓	✓	✓	✓		✓	☑	✓	✓	✓	✓		✓
Doubly incontinent					✓		✓		✓	✓				✓	✓	✓	✓			
Communication																				
Unable to speak										✓	✓				✓	✓	✓			
Speaking less/not initiating	✓	☑	✓		✓	✓			✓	✓	✓	✓	✓	✓	✓	✓	✓		✓	✓
Slow, slurred speech, not finding words	✓	✓	✓	✓	✓				✓		✓	☑	✓	✓	✓	✓	✓			
Loss of reading and writing skills	✓	✓	✓	✓				✓		✓	✓			✓		✓				
Verbal noises					✓		✓		✓	✓	✓				✓	✓	✓			
Unable to follow instructions	✓	✓	✓	☑		✓		✓	✓	✓	✓		✓							
Coordination, orientation and independence																				
Losing track of time	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓			✓			✓	✓
Restlessness, agitation											☑						✓			
Changed sleep pattern	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓	✓
Wandering (at night/ indoors/out of doors)	☑	✓					✓	✓					✓	✓			☑	✓	✓	✓
Not able to find way in familiar surroundings			✓	✓					✓			✓	✓		☑	☑	✓	✓	☑	
Unable to handle money			✓									✓								
Unsteady, shuffling walk, crawling	✓	✓	✓	✓	✓				✓		✓		✓				✓	✓		
Falls			✓					✓	✓	✓			✓			✓				
Fits (epilepsy)				✓			✓		✓	✓	✓		✓	✓						✓
Unable to walk										✓					✓	✓	✓			

Table I Continued

Symptoms	101	102	104	105	106	107	108	109	110	114	115	116	117	118	120	122	123	125	128
Unable to recognize risks and avoid dangerous situations	✓		✓																
Memory problems and social behaviour																			
General forgetfulness and confusion	☑	✓	☑	☑	✓	☑	✓	✓	✓	✓	✓	✓	☑	☑	✓	✓	✓	✓	☑
Unable to recognize familiar persons (staff/relatives)	✓	✓	✓	✓	☑		✓	✓	✓	✓	☑		✓	✓	✓	✓	✓		
Unable to remember recent events	✓	✓	✓	✓	✓	✓		✓	✓		✓	✓	✓	✓	✓				✓
Able to remember past events/speaking of past as present	✓	✓	✓	✓	✓									✓					✓
Withdrawal from social activities and/or persons/loss of interests/hobbies	✓	☑	✓	✓	✓	✓	✓	✓	☑		☑	✓	✓	✓	✓	✓	✓	✓	✓
Obsessive behaviour	✓	✓					✓	✓						✓		✓		✓	✓
Verbal aggression				✓	✓	✓	✓	✓	✓	✓	✓		✓	✓					
Physical aggression			✓			✓	✓	✓		✓				✓					
Suspiciousness and accusations				✓															✓
Hoarding and hiding/possessiveness		✓				☑	✓	✓											✓
Psychological																			
Hallucinations/delusions	✓		✓		✓			✓						✓					
Talking to self	✓		✓		✓			✓		✓									
Insecurity/anxiety/nervousness	✓	✓	✓	✓	✓					✓									✓
Attention seeking	✓					✓		✓											✓
Depression/crying	✓		✓		✓		✓	✓	✓	✓	✓		✓	✓	✓			☑	
Awareness of dementia	✓		✓	✓								✓	✓	✓		✓			
Additional problems																			
Sight problems			✓		✓												✓		✓
Hearing problems			✓		✓														✓

The numbers in the top row are those allocated to each patient by M.H.; the ticks in the boxes are the symptoms described by carers specifically as early symptoms.

(5) forgets what he was going to tell you; (6) loses things because cannot remember where he put them; (7) cannot tell what he did and ate in the day centre (an example of episodic memory impairment); (8) has forgotten that his close relative has died recently (an example of explicit memory impairment); (9) cannot remember what had happened last week; (10) does not know where the telephone is (an example of spatial memory

impairment); (11) does not know which button to press on the telephone (an example of procedural memory impairment); (12) cannot remember what day of the week it is; and (13) forgets that she had something 10 min ago.

Confusion

Along with the evidence of problems with the recent memory, many adults with DS showed

Table 2 A selection of verbatim quotes from qualitative interviews using pseudonyms throughout

- I17** – ‘The first kind of things I noticed was that he was getting forgetful. Where things went, ahm and he’d go to the cupboard to get a cup and it was not that cupboard it was another cupboard. And . . . and that’s was how, that was the first signs I realized that perhaps something was going on’.
- I10** – ‘That’s when we noticed; when he went off his music we could tell then that he was deteriorating. He lost interest in his books ahm, and he . . . he always had his, when he sat here watching television he always had his, he had a train book in his lap, reading about planes and trains. He couldn’t read, just the photographs he would look at. and we used to take him to Rhoose airport to ahm, to look, ahm, to see the planes. He used to go on the top then and ahm, he loved that. Ahm, he thoroughly enjoyed it, ahm, but that’s how we noticed when he started going off his books, going off his music, getting confused when he was putting his records on’.
- I01** – ‘Ahm, and from what I understand things then rapidly started going downhill with Anna. She was out wandering the streets at night, she do things like leave newspaper on top of a lamp that was lit, you know’.
- I04** – ‘The fact that he was just getting a little forgetful. So but it got to the point then where when he did have his tablets then where he had to take one every day he would take it in the morning he’d go days and days and days he wouldn’t take them. and then he all of a sudden he’d take 3 or 4’.
- I22** – ‘She’d moved in and they were going abroad on holiday and whilst they were in Spain it was noted that she was getting up in the night ahm, wandering around the hotel. Ahm, her basic skills, such as her eating and drinking . . . I mean, she was very fussy, you know, and she used a knife and fork correctly, etc., ahm, her eating habits began to deteriorate. and this basic confusion where she was in . . . in . . . with regards to orientation really. That first triggered a few sort of suspicions, ahm, on return from the holiday . . . I mean obviously they were periodic, she would get up in the nights, ahm, things started to deteriorate, she would urinate in her bedroom, ahm, she would go through her wardrobes, she would pull clothes out, ahm, and within day services . . . I mean, I remember her coming up to me saying Where am I?, I am lost, I need to go here, I need to go there . . . whereas before I mean she would have had no problem whatsoever’.
- I18** – ‘I think yeah, wetting himself and forgetfulness. But which is hard to pick out because we all get a bit forgetful. But it was becoming marked because he would forget what he went up for. It just went . . . Ahm, I mean, there were a . . . lots and lots of things that was gradual. But I think in the end it was the aggression, wasn’t it’.
- I07** – ‘But she just don’t want to go anywhere either, does she Frank. She used to love to go to the market you know. But now she’s not interested. Unless she really wants something and then she . . . she’ll say all right I’ll go’.

The numbers in the beginning of paragraphs represent the numbers allocated to each case by M.H.

symptoms of confusion early on in the dementing process. For example, one person threw away a £5 note in the bin instead of a receipt after coming back from a shop with a receipt and a £5 note. Other examples of confusion provided by carers included references to the DS adults: (1) putting on vests and shirts the wrong way round; (2) doing the buttons the wrong way; (3) putting two legs in one trousers leg; (4) putting two socks on one foot; and (5) putting on pullover back to front.

Problem with instructions

Many adults with DS who took part in the study exhibited problems in following more than one instruction at one given time. One example was of a person who picked up all knives or all forks when he was asked to pick up a knife and a fork from a drawer in the next room.

Slowness

Many adults with DS in our study showed evidence of slowness that affected almost all aspect of their functioning including walking slowly, eating slowly, speaking slowly, particularly failing to initiate conversations, and slowness in the overall body movement.

Speech and language problems

Many adults with DS in our study showed speech and language problems in the form of tardiness in instigating a conversation, lack of expression and flattening of the tone of voice, and repetitive questioning (which was not present previously). Other examples include speech becoming slow and difficult to understand (which was not the case before), and problem with their own comprehension, which

Table 3 Examples of symptoms of dementia among adults with Down's syndrome of different genders and with different degrees of ID

	Early symptoms	Examples of other symptoms
Male, aged 59, mild ID	Incontinent of urine, general forgetfulness	Unable to wash and dress unaided, feed unaided and use toilet, soiling, speaking less, slow speech, loss of reading and writing skills, changed sleep pattern, wandering, epileptic fits, unable to recognize familiar persons, verbal aggression, crying
Male, aged 56, moderate ID	Speaking less, not initiating conversation, withdrawal from activities/persons	Unable to wash and dress unaided, unable to use eating utensils, slow speech, unable to follow instructions, changed sleep pattern, wandering, unsteady walk, forgetfulness, hoarding and hiding items, anxiety
Male, aged 72, severe ID	Unable to recognize familiar persons	Unable to wash and dress unaided, unable to feed unaided and use toilet, doubly incontinent, speaking/signing less, verbal noises, losing track of time, changed sleep pattern, unsteady walk, verbal aggression, talking to self, sight and hearing problems
Female, aged 58, mild ID	Wandering, general forgetfulness	Inappropriate dressing, overeating, speaking less, slow speech, loss of reading and writing skills, unable to follow instructions, losing track of time, unsteady walk, withdrawal from social activities, hallucinations, talking to self, insecurity/anxiety, crying
Female, aged 55, moderate ID	Stripping off/exposing, crying	Unable to wash and dress unaided, overeating, unable to use eating utensils, problems to chew and swallow, doubly incontinent, unable to speak, verbal noises, changed sleep pattern, epileptic fits, unable to walk, unable to recognize familiar persons
Female, aged 71, severe ID	Unable to recognize familiar person, withdrawal from familiar person	Unable to wash, dress, eat and use toilet, fear of water, stripping off/exposing, unable to speak, changed sleep pattern, epileptic fits, unsteady walk, crying

ID, intellectual disabilities.

was manifested in the inability to follow simple instructions.

Sleep problems

Sleep problems in the forms of early morning waking and wandering at night were prominent features in a number of adults with DS in our study. Some showed evidence of catnapping during the day at the expense of less and less sleep at night. Some were reported to be switching the bed light on and off continuously during the night, not sleeping at all during the night and instead speaking to themselves all night long.

Loss of skills

There were many examples of loss of skills among adults with DS as they developed dementia. A number of carers reported the need for more prompting and help to perform tasks that the person with DS was previously able to carry out with little or no help. Some examples of loss of skills mentioned by the carers included references to the person with DS not being able to: (1) tell the time; (2) switch on his CD player; (3) print his name; (4) do shopping on her own; (5) use the phone; and (6) write her name. Others reported that the person with DS was doing less and less for themselves, and needed a lot of help

in the bath or someone to supervise them all the time. Loss of skills was particularly prominent in early stage of dementia among adults with severe ID.

Problems with socializing

A number of carers mentioned problems with socializing among the adults with DS who had developed dementia. Examples provided by the carers include: (1) not socializing; (2) becoming reclusive; (3) going into his own world; (4) providing very little eye contact; (5) having a vacant look; (6) putting head down all the time; (7) going off into a little fantasy world of his own; (8) refusing to communicate; and (9) becoming very quiet and less sociable in general.

Lack of confidence

Some adults with DS in our study lost confidence in themselves since developing dementia. This was manifested through nervousness, fearfulness, shakiness, low self-esteem, nervous when walking on an uneven surface, seeking company when unwell, becoming fearful of height and of bridges or stairs, developing a tendency to give up easily, lacking self-confidence and worrying excessively about everything.

Loss of interest

A number of adults showed a lack of interest in life, which was manifested through the lack of motivation and behaviour perceived as laziness. Some carers reported that they could not get people to do things because the adults with DS did not want to try things. This was not always associated with depressed mood; in fact, mostly these symptoms were associated with apathy and social withdrawal. Loss of interest in most cases were pervasive in nature, including the loss of interest in food, watching drama, going out, watching TV, going to discos, interest in friends and family members, and socialization in general.

Obsession

Some adults with DS developed what appeared as obsessive symptoms. For example, they might take excessively long time to complete a task such as

eating. Similarly, they may repeat the same movements many times or watch the same videotape over and over again or go to the toilet several times.

Balance problems

Balance problems were not uncommon among the adults with DS in our study. Many developed unsteady gait, became fearful of kerbs, started to lean over to one side and shuffle. In some extreme cases, the adults with DS refused to walk altogether, slumped on the chair and did not sit straight. There were reports of people falling off a chair or a bed, and problems climbing stairs (which could be managed before).

Emotional problems

Emotional problems such as crying episodes, becoming easily tearful, getting upset about parents who had died a long time ago and thinking that they were still alive, getting easily upset and frustrated, and shouting and screaming were reported in a number of adults with DS in our study.

Hallucinations and illusions

Some carers reported sensory problems in the form of illusions and hallucinations among adults with DS that they cared for. Examples of hallucinations include reporting of seeing things that are not there (visual hallucinations). One such example involves the report of an adult with DS who felt that a cat was rubbing against his body while in fact the cat was sitting in the same room at a distance. This experience may be interpreted as tactile hallucinations (feeling of touch-related sensation in the absence of an external stimulus). A similar example involves a report of a person who picked up things (imaginary) out of the air.

Covering up

A number of carers reported an interesting phenomenon among adults with DS who developed dementia, which we have presented here under the heading 'Covering up for the loss of memory'. Many adults with DS constantly kept saying 'sorry' when they could not remember things. Some disguised things that they lost and some frankly

confabulated (made up stories to cover up for their lost memory).

Hypochondriasis

Some carers reported that since developing dementia some adults with DS became what they called 'hypochondriac', in that they reported many bodily symptoms such as 'tummy ache' for which no obvious physical cause could be found. Such behaviours were reported to be 'out of character'.

Change in personality

Some carers reported a change in personality since the person with DS had developed dementia. Often change in personality in this context means the person becoming aggressive and behaviourally disturbed. In the current study, however, the carers reported a different type of personality change. For example, a person with DS might be reported to have lost his previous sense of mischievousness rather than becoming aggressive.

Case studies

We have described below two cases in order to demonstrate differential manifestation of dementia symptoms among adults with mild and severe ID.

Case 1: M.S. was a man with mild ID who was living independently with minimum support and had no physical problems. Prior to developing dementia he was described as articulate, literate, sociable and polite with good sense of humour. He developed dementia and died from pneumonia at the age of 55 years. His carers reported first symptoms of dementia as forgetfulness, anxiety and loss of skills such as the ability to handle money.

Case 2: M.S. was a man with severe ID who lived in a nursing home. He had multiple health problems. He was also deaf and blind. He developed dementia and died of pneumonia at the age of 58 years. His carers described first symptoms of dementia as uncharacteristic social withdrawal, verbal and physical aggression, changed sleep pattern and loss of skills such as feeding.

Discussion

Our study contains the first detailed description of dementia symptoms, particularly the early symp-

toms based on direct accounts of carers of adults with DS who had developed dementia, rather than on clinician's reports. The importance of qualitative methodology in health service research and more particularly in dementia research has been emphasized by many authors (see Murphy *et al.* 1998; Gibson *et al.* 2004). Only recently qualitative methodology has been employed to develop questionnaires (see Espie *et al.* 2001; Morris *et al.* 2005) for patients with brain damage.

It is interesting to note that most early and primary symptoms of dementia among adults with DS in our study were similar to those found in adults without ID. For example, the loss of recent memory in the absence of loss of distant memory was the early prominent symptom in most cases of adults with DS. This trend was particularly noticeable among adults with a mild-to-moderate degree of ID, as depicted in the case reports in the Results section. This is also the case for people without ID. However, these symptoms may be difficult to detect, particularly in the early stage of dementia among adults who have severe and profound ID. This has been demonstrated by the case reports presented in the Results section. In these people, loss of skills and some behavioural problems seem to be the early indicators of dementia. It is important to point out here that the early symptoms in this context are determined from the narratives of the carers (i.e. what the carers described in the interview as the symptoms that they have noticed first). However, carers' memory may not be totally accurate in this context when they were describing events retrospectively. Therefore, a true account of the order of appearance of different symptoms could only be determined from a prospective follow-up study of the same cohort from the onset of dementia up to their death. Similarly, we could not determine the relative weighting of these symptoms, therefore reported only the frequency at which these symptoms were described by the carers.

It has been shown in the general population that different aspects of memory deficits appear at different stages of the dementing process. For example, explicit memory is affected long before the implicit memory. Similarly, the recent memory is affected earlier than the distant memory. This was evident in our study. Although we tried to describe certain types of memory deficits in the

Results section, the exact nature of memory deficit could only be determined by using a battery of memory-specific neuropsychological tests. For example, episodic memory deficit is traditionally assessed using the 'Story Recall Test'. Most screening instruments used in the general population, for example, the MMSE, do not distinguish among different types of memory impairments. As our aim was to develop a screening questionnaire based on carers' reporting, it was not possible to differentiate among various aspects of memory deficit within this study. This should be determined by using specific memory tests. It is important to highlight here that there is a dearth of validated neuropsychological instruments for use among people with ID, which will differentiate among different types of memory deficits.

Many early symptoms described by the carers such as 'social withdrawal', 'pervasive slowness', 'diminished initiative', etc. point more towards the features of the 'frontal lobe dysfunction' or 'executive dysfunction'. These symptoms are usually reported at a later stage of dementia in the general population. Holland *et al.* (1998) reported features similar to those associated with fronto-temporal dementia as early/preclinical presentation in a subgroup of adults with DS in their study group. As a possible explanation, the authors suggested that these findings are due to an interaction between Alzheimer's neuropathology and the altered brain structure in adults with DS, but not due to distinct neuropathology of fronto-temporal dementia. In a prospective follow-up of Holland *et al.*'s (1998) cohort, Ball *et al.* (2006) reported that 'the early presentation of Alzheimer's disease in people with DS is characterized by prominent personality and behavioural changes, associated with executive dysfunction, providing support that the functions of the frontal lobes may be compromised early in the course of the disease in this population'.

We believe that there are two possible explanations for finding such prominent features of 'frontal lobe' dysfunction early in dementia among adults with DS. First, people with DS show structural abnormalities in the frontal and the temporal lobes (Deb 1997), and are therefore vulnerable to loss of frontal and temporal lobe functions early because of any brain insult. Second, it is often not possible to detect dementia in its early stage among adults who

have ID. Therefore, dementia tends only to be diagnosed in these people when the disease has already progressed to its middle or late stage. This is also evident from the shorter period of time that is clinically observed between making the diagnosis of dementia and death among adults with DS compared with people without ID. The features of 'frontal lobe' dysfunction are not uncommon in the later stage dementia in the general population; therefore, it is not surprising to detect these features fairly early in the diagnosis among adults with ID, given that these people are probably already in an advanced stage of dementia by the time a diagnosis is made.

Symptoms associated with confusion such as putting two legs in one trousers' leg may have been the manifestation of disorientation or memory problem or dressing dyspraxia. Similarly, slowness of speech and failure to initiate conversation may be explained by either language problems or apathy symptoms, which are associated with the frontal lobe dysfunction. Obsessive tendencies may reflect the development of true obsession or may have been caused by loss of memory for which the person needs to check everything several times or may be the part of overall slowness in information processing or perseveration caused by frontal lobe dysfunction. Similarly, failure to carry out more than one instruction at a time may have been caused by the loss of procedural memory or language difficulty such as comprehension problems or slowness in information processing, which again is associated with frontal lobe dysfunction.

The hallucinations reported were primarily 'visual' in nature as opposed to 'auditory', which are more characteristic of functional psychiatric illness such as psychoses. Frank 'delusions' were not reported, although there was one report of an adult complaining of a cat rubbing against him while the cat was in fact sitting at a distance. This could be a delusion or a tactile hallucination or pure imagination.

We have found a differential manifestation of dementia in the early stage depending on the severity of ID. However, Cosgrave *et al.* (2000) reported memory loss as early clinical feature of dementia among 85.7% of both the 14 females with moderate and seven with severe ID. Spatial disorientation was the next most frequent symptom and was present in

both those with moderate and those with severe ID. Epilepsy and urinary incontinence were more commonly present at diagnosis of dementia in the severely intellectually disabled group than in the moderately intellectually disabled group, but apathy symptoms were more common among adult females with moderate ID than among those with severe ID. None of these differences were statistically significant though.

In the general population, carers of people with dementia find that among the non-cognitive symptoms, aggression is one of the common causes of burden for care (Donaldson *et al.* 1997). However, in general a lower rate of aggression is reported among people with DS either with or without dementia (Collacott *et al.* 1998). In a study of 128 adults with DS, 29 had dementia; Cosgrave *et al.* (1999) found that the presence of dementia was not predictive of aggression. Prasher & Filer (1995) also recorded a low prevalence of significant maladaptive behaviour in people with dementia and DS. In a study of 19 adults with DS and dementia, and 26 adults with other causes of ID and dementia, Cooper & Prasher (1998) found that aggression occurred with greater frequency in those subjects with ID due to other causes, but low mood, restlessness/excessive overactivity, disturbed sleep, uncooperativeness and auditory hallucinations were more common among adults with DS and dementia. Huxley *et al.* (2005) found a higher rate of challenging behaviours among 15 adults with DS and dementia compared with 19 adults with DS who did not have dementia. However, in their study challenging behaviour did not include aggression, but included behaviours such as irritability, hyperactivity, stereotypy and lethargy.

The family carers in the current study did not report severe behavioural problems, although they were not altogether absent. As our study analysed data based on carers' report, it is possible that the carers may have underreported the behavioural symptoms. We believe that some of the cognitive deficits such as language problems or inability to follow instructions may have indirectly caused behaviour problems through frustration. Similarly, the lack of initiative, which was sometimes interpreted as sheer 'laziness', may have caused frustration and led to aggression. Similarly, if someone has lost interest in activities and has become

socially withdrawn, any demand placed on that person in order to comply with social activities is likely to cause further friction, which may lead to aggression. We believe it is of utmost importance to try to find an explanation and an underlying cause for aggression in a person who has developed dementia in order to minimize unnecessary use of medication.

In the current study we have not used an age-matched control group of people with DS who did not develop dementia; therefore, it may be difficult to say that all the behavioural changes reported by the carers are associated with dementia rather than the normal ageing process. However, considerable debate exists regarding the overlap of age-related cognitive decline and dementia in the general population as well as in people with ID. Studies that have mapped out cognitive decline among adults with ID (with and without DS) using prospective design (Devenny *et al.* 2000; Carr 2005) show a trend of relatively greater decline in verbal abilities compared with performance abilities. However, the exact features associated with normal ageing and their overlap with dementia symptoms have never been properly studied among people with ID.

It is imperative to make an early diagnosis of dementia so that an early treatment can be instigated and appropriate planning for service provision can be made at the right time. Hypothyroidism and depression are common in people with DS and can mimic dementia. It is important to differentiate between these conditions and dementia, as these conditions are often treatable. A false positive diagnosis of dementia has an equally devastating effect on patients and their families as a false negative one. The qualitative study described here provides the basis for the development of a screening questionnaire; the development and validation of this measure is described in a different paper (see Deb *et al.* 2007).

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References

- Aylward E. H., Burt D. B., Thorpe L. U. & Dalton A. J. (1997) Diagnosis of dementia in individuals with intellectual disability. *Journal of Intellectual Disability Research* **41**, 152–64.
- Ball S. L., Holland A. J., Hon J., Huppert F. A., Treppner P. & Watson P. C. (2006) Personality and behaviour changes mark the early stages of Alzheimer's disease in adults with Down's syndrome: findings from a prospective population-based study. *International Journal of Geriatric Psychiatry* **21**, 661–73.
- Brugge K. L., Nichols S. L., Salmon D. P., Hill L. R., Delis D. C., Aaron L. & Trauner D. A. (1994) Cognitive impairment in adults with Down's syndrome: similarities to early cognitive changes in Alzheimer's disease. *Neurology* **44**, 232–8.
- Carr J. (2005) Stability and change in cognitive ability over the life span: a comparison of populations with and without Down's syndrome. *Journal of Intellectual Disability Research* **49**, 915–28.
- Collacott R. A., Cooper S. A., Branford D. & McGrother C. (1998) Behaviour phenotype for Down's syndrome. *British Journal of Psychiatry* **172**, 85–9.
- Cooper S.-A. & Prasher V. P. (1998) Maladaptive behaviours and symptoms of dementia in adults with Down's syndrome compared with adults with intellectual disability of other aetiologies. *Journal of Intellectual Disability Research* **42**, 293–300.
- Cosgrave M. P., Tyrell J., McCarron M., Gill M. & Lawlor B. A. (1999) Determinants of aggression, and adaptive and maladaptive behaviour in older people with Down's syndrome with and without dementia. *Journal of Intellectual Disability Research* **43**, 393–9.
- Cosgrave M. P., Tyrell J., McCarron M., Gill M. & Lawlor B. A. (2000) A five year follow-up study of dementia in persons with Down's syndrome: early symptoms and patterns of deterioration. *Irish Journal of Psychological Medicine* **17**, 5–11.
- Deb S. (1997) Structural neuroimaging in mental retardation. *British Journal of Psychiatry* **171**, 417–19.
- Deb S. & Braganza J. (1999) Comparison of rating scales for the diagnosis of dementia in adults with Down's syndrome. *Journal of Intellectual Disability Research* **43**, 400–7.
- Deb S., de Silva P. N., Gemmell H. G., Besson J. A., Smith F. W. & Ebmeier K. P. (1992) Alzheimer's disease in adults with Down's syndrome: the relationship between regional cerebral blood flow and dementia. *Acta Psychiatrica Scandinavica* **86**, 340–5.
- Deb S., Hare M., Prior L. & Bhaumik S. (2007) Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID). *British Journal of Psychiatry* **190**, 440–4.
- Department of Health (2006) *Supporting People with Long Term Conditions to Self Care – A Guide to Developing Local Strategies and Good Practice*. DoH, London.
- Devenny D. A., Krinsky-Hale S. J., Sersen G. & Silverman W. P. (2000) Sequence of cognitive decline in adults with Down's syndrome. *Journal of Intellectual Disability Research* **44**, 654–65.
- Donaldson C., Tarrrier N. & Burns A. (1997) The impact of the symptoms of dementia on caregivers. *British Journal of Psychiatry* **170**, 62–8.
- Espie C. A., Watkins J., Duncan R., Espie A., Sterrick M., Brodie M. J., McGarvey C. & Curtice L. (2001) Development and validation of the Glasgow Epilepsy Outcome Scale (GEOS): a new instrument for measuring concerns about epilepsy in people with mental retardation. *Epilepsia* **42**, 1043–51.
- Evenhuis H. M. (1990) The natural history of dementia in Down's syndrome. *Archives of Neurology* **47**, 263–7.
- Folstein M. F., Folstein S. E. & McHugh P. R. (1975) 'Mini Mental State'. A practical method for grading the cognitive state of patients for the clinician. *Journal of Psychiatric Research* **12**, 189–98.
- Gibson G., Timlin A., Curran S. & Wattis J. (2004) The scope for qualitative methods in research and clinical trails in dementia. *Age and Ageing* **33**, 422–6.
- Hesse-Biber S. & Leavy P. (2006) *The Practice of Qualitative Research*. Sage Publications, Thousand Oaks, CA.
- Holland A. J., Hon J., Huppert F. A., Stevens F. & Watson P. (1998) Population-based study of the prevalence and presentation of dementia in adults with Down's syndrome. *British Journal of Psychiatry* **172**, 493–8.
- Holland A. J., Hon J., Huppert F. A. & Stevens F. (2000) Incidence and course of dementia in people with Down's syndrome: findings from a population-based study. *Journal of Intellectual Disability Research* **44**, 138–46.
- Huxley A., Van-Schaik P. & Witts P. (2005) A comparison of challenging behaviour in an adult group with Down's syndrome and dementia compared with an adult Down's syndrome group without dementia. *British Journal of Learning Disabilities* **3**, 188–93.
- Lai F. & Williams R. S. (1989) A prospective study of Alzheimer's disease in Down's syndrome. *Archives of Neurology* **46**, 849–53.
- Mann D. M. A. (1988) Alzheimer's disease and Down's syndrome. *Histopathology* **13**, 125–37.
- Morris P. G., Prior L., Deb S., Lewis G., Mayle W., Burrow C. & Bryant E. (2005) Patients' views on outcome following head injury: a qualitative study. *BMC Family Practice* **6**, 1–6.

- Murphy E., Dingwall R., Greatbatch D., Parker S. & Watson P. (1998) Qualitative research in health technology assessment. *Health Technology Assessment* **2**, 1–276.
- Oliver C., Crayton L., Holland A., Hall S. & Bradbury J. (1998) A four year prospective study of age related cognitive change in adults with Down's syndrome. *Psychological Medicine* **28**, 1365–77.
- Prasher V. P. (1995a) Age-specific prevalence, thyroid dysfunction and depressive symptomatology in adults with Down's syndrome and dementia. *International Journal of Geriatric Psychiatry* **10**, 25–31.
- Prasher V. P. (1995b) End-stage dementia in adults with Down's syndrome. *International Journal of Geriatric Psychiatry* **10**, 1067–9.
- Prasher V. & Filer A. (1995) Behavioural disturbance in people with Down's syndrome and dementia. *Journal of Intellectual Disability Research* **39**, 432–46.
- Richards L. (1999) *Using Nvivo in Qualitative Research*. Sage Publications, London.
- Roeden J. M. & Zitman F. G. (1995) Ageing in adults with Down's syndrome in institutionally based and community-based residences. *Journal of Intellectual Disability Research* **39**, 399–407.
- Strauss A. & Corbin J. (1990) *Basics of Qualitative Research: Grounded Theory Procedures and Techniques*. Sage Publications, London.
- Thase M. E., Tigner R., Smeltzer D. & Liss L. (1984) Age-related neuropsychological deficits in Down's syndrome. *Biological Psychiatry* **19**, 571–85.
- World Health Organization (WHO) (1992) *The ICD-10 Classification of Mental and Behavioural Disorders: Clinical Descriptions and Diagnostic Guidelines*. WHO, Geneva.

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