

# Down's Syndrome and dementia: A service evaluation

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## Introduction

Down's Syndrome is identified as the aetiological cause of learning disabilities in 20 per cent of adults with moderate to severe learning difficulties (Cooper, 1999). Individuals with Down's syndrome (DS) have an increased risk of developing dementia, with over 50 per cent developing Alzheimer's disease (AD) before the age of seventy (Holland, 1998, Prasher, 1995). Alzheimer's disease is a dementia that includes impairment in and eventual loss of cognitive and adaptive skills necessary for successful personal, community and occupational functioning (NIH, 1987). In the earliest stages, mild memory and language disturbances occur. This is followed by more severe cognitive decline including: perceptual disturbances; loss of self care; sleep disturbance; wandering; aggression; irritability; aphasia; gait deterioration; incontinence and the development of psychotic phenomena, and seizures. Eventually individuals require total bed care and death soon follows (McKenzie, *et al.*, 2000). There is evidence that the progression of AD is rapid among people with DS with a course of 3–5 years between diagnosis and death (Kerr, 1998, Thompson, 2000), making a strong case for early recognition.

Recent advances in healthcare have meant that the life expectancy of people with DS is increasing and 70 per cent will live beyond 70 years (Baird & Sadovnick, 1987). Consequently, since the occurrence of AD is correlated with age, learning disability services are seeing an increased number of referrals of people with suspected dementia (Prasher & Krishan, 1993). This steep rise and the difficulties in assessing this population have created huge challenges for service provision (Kerr, 1998).

The diagnosis of dementia in people with DS is often problematic, especially in the early stages (Oliver, 1998, Aylward, *et al.*, 1997. McKenzie *et al.*, 2000). It is difficult to assume baseline function for the DS population, which contains wide individual differences in pre-morbid ability. Without baseline data diagnosis is often only confidently made in the late stages of AD (Duggan *et al.*, 1999). Lifestyle differences between people with learning difficulties and the general population (Cooper, 1999), and communication difficulties provide further obstacles to picking up early signs of dementia. Often poor performance is simply attributed as integral to the original learning disability.

There is no widely accepted gold standard for establishing dementia in an individual with DS (Prasher, 1997; Burt & Aylward, 1998). Cognitive and behavioural assessment tools that are often used to assess dementia in the general population are of limited value with people with learning difficulties, as they rely on good verbal skills and the absence of sensory impairments. Furthermore, cut-off scores and norms are in reference to the general population. Although a uniform battery of tests to measure clinical change indicative of dementia in DS is not yet available, there are several tools which have either been adapted or developed for learning disabled populations.

This study is an evaluation of the service currently offered to individuals with DS by a multi-disciplinary community learning disabilities team (CLDT) in the Southwest region (UK). Planning meetings identified that the team currently have an *ad hoc*, reactive response to assessing dementia in people with DS, and operate on a referral-by-referral basis. There was concern about the difficulties of assessing decline in service-users for which there was no availability of baseline data indicating previous level of function. Furthermore clinical uncertainty about the best tools to use was hindering early diagnosis with consequences for the team's ability to provide psychological intervention, pharmacological treatment and palliative care. Literature on best practice suggests that a systematic assessment procedure is desirable, and moreover that instead of waiting for cases of suspected dementia to be referred, there should be a proactive

screening system implemented with younger adults with DS (Janicki *et al.*, 1996, McKenzie, 2000).

The aims of this evaluation were therefore:

- To establish the parameters of current practice with the team for assessing dementia in people with Down's syndrome.
- To identify the service models and assessment tools used by other teams in the Southwest region.
- To examine whether there is a need for a screening system to be implemented.
- To explore the prerequisites needed for implementation and any barriers that might exist.

## Methods

Two methods<sup>1</sup> were used to:

- a) Evaluate current practice in relation to DS and dementia
  - b) Investigate the need for a dementia screening programme
- a) Audit: An audit of multidisciplinary clinical files was undertaken to establish the number of people with Down's syndrome on the current caseload. A retrospective examination of cases with diagnosed dementia allowed the trajectory of the care they received to be mapped, providing an outline of current patterns of service provision.
- b) Questionnaires: An internal questionnaire was developed to elicit: team perceptions of current practice; information about the range of assessment tools used; their views on the need for screening and any barriers or ethical concerns. A similar questionnaire was sent to external teams to identify the service models and assessment tools used by other services in the Southwest region.

## *Participants*

The internal questionnaire was distributed to representatives of each discipline (n=7): Psychology, Occupational Therapy (OT), Nursing, Psychiatry, Speech and Language Therapy (SLT), Physiotherapy,

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<sup>1</sup> The service evaluation required no direct contact with service-users and was approved by trust committee for ethical approval without a formal application.

Healthcare Assistants. The external questionnaire was sent to clinical psychologists working in learning disability teams in the Southwest region (n=30).

### *Procedure*

#### a) Audit

A comprehensive audit of multidisciplinary clinical files for the entire service caseload (n=649) was completed in March 2003. A record was made of individuals with a diagnosis of Down's syndrome and a computer database compiled. Where a diagnosis or suspicion of dementia was recorded in the notes, in-depth scrutiny of both multidisciplinary and medical notes for each individual enabled the mapping of the trajectory of care received from the point of referral for dementia assessment to the time of the audit. Dates of assessment, intervention and the professionals seen were recorded.

#### b) Questionnaires

The internal team questionnaire was developed with reference to best practice literature. It was discussed with two experts in the field of Down syndrome and dementia to ensure that pertinent issues had not been missed. The questionnaire was piloted with two members of the team. It was distributed to representatives of each discipline with an explanatory letter. Each representative was asked to liaise with all members of their discipline to provide a single response for their department.

The external questionnaire was an adapted version of the internal questionnaire and almost identical in content. The questionnaire was mailed with a covering letter and stamped addressed envelope to clinical psychologists working in learning disability services within the Southwest region. Thirty individuals were identified from the list of supervisors for the Southwest Training Scheme Doctorate in Clinical Psychology and may not represent a comprehensive survey of all teams in the region. A month was allowed for questionnaire return and no reminders were sent.

## Results

### a) Audit

Thirty-seven individuals with Down's syndrome were identified. This represents 5.7 per cent of the entire service caseload. Twelve individuals (37 per cent of those with DS) had been referred for assessment due to suspected dementia. The mean age of this group at the time of the audit was 48.8 years (range 30–66, s.d. 10.8). The group included 9 men and 3 women.

The initial assessment was most frequently conducted by a psychiatrist, who assessed mental state and health checks (blood, liver & thyroid tests), with subsequent functional assessment most often carried out by an occupational therapist. Baseline data were not automatically collected soon after referral for suspected decline, indeed such assessment was only recorded in three of twelve cases. These cases were relatively recent, suggesting that practice had changed within the last year. A further two cases had only received baseline assessment following a re-referral for more serious decline or a diagnosis of dementia. Data were then collected to monitor rate of deterioration. Baseline assessments of adaptive behaviour were conducted by an occupational therapist, tests of cognitive function by a psychologist and language assessment by a speech and language therapist. Of the twelve individuals referred for assessment, three had since received a definite diagnosis of Alzheimer's disease. The mean age at diagnosis was 55.3 years (range 53–58, s.d. 2.5). All three were diagnosed at the mid to late stage of dementia.

The care trajectories for all three individuals with dementia showed the following similarities: psychiatrists were the initial assessors; health checks were conducted; baseline function was not assessed immediately making subsequent decline difficult to chart; diagnosis was based on negative health checks, carer reports and behavioural observations and occurred at mid-late stages of dementia. After diagnosis there was no consistent package of care or therapeutic

intervention offered to individuals and care management was largely taken over by social services.

#### b) Questionnaires

Seven internal questionnaires were returned. Six of seven disciplines responded. A further two individual responses were received from a psychiatrist and a nurse. Of the thirty questionnaires sent to clinical psychologists working in other teams, twelve were returned giving a response rate of 40 per cent.

#### Current Service

All eight respondents from the team stated that assessment is always conducted on a case-by-case basis and that there was no fixed procedure. This mirrored the services operated by five of twelve regional teams. A further five regional teams reported having a fixed procedure, although this was not always followed (3). One team reported having established a screening programme for dementia although this had recently been abandoned. There was near unanimous agreement within the team that the referral was not automatically passed to a single person or discipline and that multiple professions were involved in assessments. Of the other regional teams six stated that the referral would be passed to an identified discipline. Clinical psychologists were most often named (3), followed by Psychiatry (2) and Community Nursing (1).

There was a consensus on the range of methods routinely used across both surveys. These included: client and carer interviews; direct observation; assessment of current cognitive function and repeat of previously administered cognitive tests; repeat of previous language and functional assessments; assessment of general health and elimination of other reasons for decline. Several disciplines at the team reported using assessment tools (see appendix 5 for overview). Direct cognitive assessments were not used to obtain diagnostic cut-off scores, except for the Mini-mental state examination (MMSE, Folstein *et al.*, 1975), which was occasionally used by psychiatrists. Some other regional teams did use general dementia assessments such as

Dementia Rating Scale (DRS) (2) and Middlesex Elderly Assessment of Mental State (MEAMS) (2).

Both questionnaires yielded similar data about the stage of dementia at referral. Figure 1 shows that half the respondents from the team believed that referral occurred during the middle stages (4). Only one discipline thought that referrals were made early. Two believed referral occurred at a late stage and one was undecided. A similar pattern was obtained for teams across the region, suggesting that poor carer and referrer awareness of early signs of dementia may be a contributing factor.

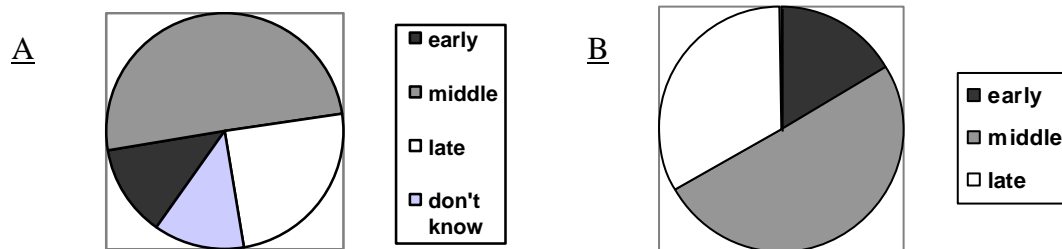


Figure 1: Responses to the question: 'Do service users with dementia usually get referred for assessment during the early, middle or late stage of dementia?' for A) CLDT and B) Psychologists from other CLDTs in SW region.

### Is there a need for dementia screening?

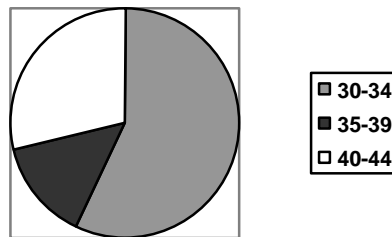
There was a strong agreement at the team that dementia screening is needed (7), with only one individual stating that they did not know. The qualitative reasons given were:

- To enable collection of baseline assessments (4)
- To facilitate earlier diagnosis (4)
- Because of known increase risk of dementia in DS population (3)
- To enable earlier carer support (3)
- Early intervention to maintain skills and improve quality of life (2)
- Earlier treatment with anti-dementia medication (1)
- To monitor change from baseline (1)
- To assist in planning and co-ordination of care (1)

Among other teams, the consensus was less strong. Seven respondents believed that there was a need for screening of dementia and their reasons were very similar to those elicited from the internal questionnaire. The following reasons were given by those answering 'no' (4) or don't know (1).

- Uncertainly about service users' ability to consent (2)
- Impractical to set up
- Value of screening is dubious because tools are rarely sensitive enough to pick up early changes
- Anti-dementia vaccines will soon be developed

Only one team had implemented a screening service. This had involved the collection of baseline measures of cognitive function by the clinical psychologist for all service-users with DS on the caseload (18 years+). The baseline screen has now been suspended by this service due to over-reliance on the time of one professional.



**Figure 2: Team responses to the question, 'At what age should individuals be invited to take part in a screening programme?'**

Figure 2 shows that the majority of disciplines at the team believed that screening should be offered to individuals from the age of 30–34 upwards. There was also clarity about what information should be collected at the initial baseline screen, which included:

- Baseline assessment of adaptive function (6)
- Baseline assessment of cognitive function (6)
- Assessment of physical health (5)

Responses were more divided over the model for implementing screening. Disagreement within disciplines was reported. Five respondents indicated that individuals should subsequently be monitored for decline from baseline but three stated that baseline data should be stored until concern is raised.

### Implementation of a screening service

Respondents were asked to list the factors they considered necessary for the successful implementation of a screening service. The categories of responses are listed in table 1.



Factor	No.of Respondents
Clear and consistent process/ care pathway	7
Resource allocation: staff, time, finance	6
Admin support/ keeping accurate records and database	5
Consent and compliance of service users	4
GP awareness and commitment	3
Organisational and managerial support	3
Multidisciplinary involvement and commitment	3
Agreed standardised assessment tool/ checklists	2
Tied in with other health screening	1

**Table 1: Factors for the successful implementation of a screening service**

The absence of these factors was also named as a possible barrier to successful implementation of screening. Other identified barriers included: a lack of well researched validated assessment tools (2), a lack of named co-ordinator/lead discipline (2), one discipline working in isolation (1), a lack of opportunity to review (1) and poor uptake (1). There was strong agreement that screening would involve collaboration with other agencies (7). Primary care GPs and practice nurses were most often identified (6), followed by social services (3). Other agencies mentioned included dementia specialists, older adult services, day care services and housing associations.

### Ethical issues

Three respondents reported having no concerns about consent or ethical issues stating that screening, even where informed consent could not be obtained, would be in the best interests of service-users. Five respondents expressed concerns, which were categorised and are shown in table 2.

Concern	No.of respondents
Inability to give informed consent	4
Lack of follow up, intervention or care services	4
Danger of misdiagnosis/over diagnosis	1
Stigmatising experience if screened too often	1
Lack of methods for explaining screening and dementia to service users	1
Screening must obtain approval by trust ethics committee	1

**Table 2: Ethical Concerns identified by questionnaire respondents at the CLDT.**

## Discussion and Conclusions

Current practice operates on an *ad hoc* basis, with no clearly defined procedure or care pathway. Initial assessments and health checks are usually led by psychiatry with subsequent input by other professions. This stands in contrast to the MDT's perception that there was no lead discipline. Diagnosis occurred at a mid-to-late stage of dementia. The reasons for this may be a lack of carer awareness about early signs of dementia resulting in delayed referral, and/or a lack of measurements of baseline function. The pattern of delayed diagnosis for individuals with suspected early dementia (where it was later confirmed) is indicative of clinical uncertainty. This uncertainty extends to the use of assessment tools. Diagnosis was rarely made on the basis of an explicit measure of decrement in function, but rather from carer reports, behaviour observation and the elimination of other reasons for decline.

As a team, measures of baseline function were preferred to diagnostic cognitive assessments, which are of dubious validity with this population. However, baseline assessment was not automatically initiated if decline was suspected. Instead, data were often only collected after diagnosis as a means of monitoring deterioration. After diagnosis there was no consistent package of care, therapeutic or psychological intervention and little evidence of multi-agency working. Indeed service input seemed to diminish once diagnosis had been made and the case was passed to social services for a care needs assessment. This stands in contrast to the literature on best practice, which suggests there is an ongoing role for CLDTs after diagnosis (Aylward *et al.*, 1997, Burt & Aylward, 1998, Janicki, *et al.*, 1996, McKenzie, 2000; Oliver, 1998).

There was clear agreement within the team that dementia screening is desirable for individuals with DS from around 30 years of age to facilitate earlier diagnosis and intervention. The team agreed that the most useful baseline measures would include cognitive, behavioural and health function. If implemented, this procedure would mirror current guidelines for best practice (Aylward *et al.*, 1997, Burt &

Aylward, 1998, Janicki, *et al.*, 1996, McKenzie, 2000, Oliver, 1998). The guidelines also suggest that screening should be repeated regularly to ensure that dementia is picked up at the earliest possible stage. Consensus was less clear on this issue.

## **Recommendations**

Several recommendations arise from this service evaluation.

### *Screening*

- Screening for dementia should be implemented with all service-users with DS aged 30 years or above. Ideally, screening should be repeated at annual health checks and tied in with hand held health records, to facilitate regular screenings with the least possible intrusion and to reach larger numbers of individuals with DS living in the catchment area.
- A particular effort should be made to gain support for screening from primary care teams.
- A Multidisciplinary working party should be set up to agree on screening tools. The working party should include representatives from other CLDTs within the Primary Care Trust.
- All disciplines should be involved in an in-depth discussion of ethical issues and any negative impact on service-users.
- A simple and accessible procedure for obtaining informed consent should be developed. There should be team agreement about the procedure to follow when an individual is unable to give informed consent.
- The screening programme must meet the approval of the PCT and ethics committee.
- A computer database of known individuals with DS has been compiled to facilitate screening; this should be maintained and kept up to date.
- Once established, the screening programme should be monitored and reviewed.
- In the interim, awareness training about the early signs of dementia could be given to residential staff, day care teams and carers, to encourage earlier referral.

### *Care pathway*

- A screening programme should be one element of an integrated care pathway. This should be developed in collaboration with all multi-agency stakeholders.
- Links should be developed with older adult services and voluntary dementia organisations.
- Priority should be given to developing interventions and a package of support for those diagnosed with dementia and their families.
- An accessible, pictorial information pack about dementia should be developed for service-users with DS and early dementia.

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