

How should services meet the needs of people with Down Syndrome who develop dementia?

People with Down syndrome are living longer. They also develop dementia, particularly Alzheimers, much earlier than others. As a result there is a growing need to promote appropriate assessment and care planning, and to ensure that housing and support services recognise and respond to the particular needs of those individuals with Down Syndrome who are living with dementia.

What's the subject?

Although people with Down syndrome probably have the same lifetime risk of developing a Dementia of Alzheimer type (DAT) as the rest of the population (Holland et al, 1998), this occurs 30-40 years earlier. About 22% of those with Down Syndrome in their 40s or older and 56% of those age 60 or older will develop DAT (Janicki and Dalton, 2000). This has become an issue as people with Down Syndrome now have an average life expectancy of 50 years (Holland, 1999). Down Syndrome is the commonest known cause of mild and severe intellectual disability (Minns, 1997) and may account for 15-20% of those with intellectual disability (DSA, 2007). Assessment tools for dementia in the general population are not appropriate for people with intellectual disability, and generic services for older people are not usually tailored for this group, with very few services adjusting their practice accordingly. The many specific references to intellectual disability issues in the recent NICE/SCIE guideline for dementia (2006) reflect an emerging consensus regarding best practice in this area and emphasise the importance of joint agency working. To date, few services in the UK have adjusted their practice to meet this new challenge.

Why is it important to service users?

If social care and health services (learning disability and/or older people's services) are not adapted for the needs of people with intellectual disability, especially those with Down Syndrome who develop dementia, this will lead to avoidable loss of health-related quality of life as a result of non-diagnosis. Such adaptation needs to include relevant screening, assessment, diagnosis, integrated medical and social care, follow-up and attention to end-of-life issues. There is a particular social context in that, unlike other people with dementia, the parents of individuals with Down Syndrome are often still alive and in many cases there has been little

contact with services during adulthood. Lack of health screening programmes may mean early signs of dementia are missed, or attributed to other aspects of learning disability, and opportunities for potentially useful health and social care interventions are lost. The usual protocols used to assess dementia in the general population are not appropriate for people with intellectual disability. Widely-used tools such as the Mini-Mental State Examination (MMSE) assume the level of functioning prior to onset to have been mainstream. For people with Down Syndrome this is further compounded by the fact that neuro-imaging may show characteristic changes of DAT from childhood, long before the onset of any clinical dementia, rendering such tests unhelpful in diagnosis (NICE, 2006).

Specialist expertise is therefore required. Among the variety of other health problems for which people with Down Syndrome may be especially at risk in later life is hypothyroidism, a treatable condition with an insidious onset that can mimic dementia in its presentation. Two further problems common amongst people with Down Syndrome that may confuse diagnosis are sensory (eg visual or hearing) impairments, which may deteriorate with age, and depressive illness. These potentially treatable problems can present with similar symptoms to dementia, and may of course occur both independently or alongside it. Unfortunately the evidence is that all too often such disorders remain undetected in individuals with learning disabilities (see OutLine 7). Although DAT is the most common form of dementia in people with Down Syndrome, other types of dementia (eg multi-infarct) can also occur. These may require different management approaches and lead to different outcomes.

Three important implications flow from these observations.

First, identification and screening of the population at risk might aid early diagnosis and provide access to useful health and social care interventions. Second, assessment procedures should be adapted to take into account both the learning disability and the special presentation of people with Down Syndrome. Third, social care agencies must have the capacity and knowledge to respond to the changing needs of people with Down Syndrome as they age.

What works - assessment and treatment

Good practice guidance from the Foundation for People with Learning Disabilities (Turk et al, 2001) recommended that every service for people with intellectual disability should set up a register of adults with Down Syndrome; conduct a baseline assessment of cognitive and adaptive functioning before the age of 30 years; develop specialist skills in this area; offer training to other professionals, front line staff and carers; and seek high quality co-ordination between agencies. The NICE/SCIE (2006) guidelines also make reference to the training needs of staff caring for people at risk of early dementia, including those with Down Syndrome and intellectual disability.

Availability of screening and treatment across the UK is not equitable. Although in most areas reactive screening is provided for those with signs of deterioration, very few services offer prospective screening to all young adults with Down Syndrome, such as that described by McBrien et al

(2002). As risk rises with age, a two yearly screen for people in their 40s and an annual screen for those in their 50s or older will increase the chances of picking up early signs of dementia.

Screening not only requires co-ordination between primary and secondary health care, which could be achieved as part of a Health Facilitation programme (Department of Health, 2001) but good practice also requires a co-ordinated approach with social care agencies so that the individual's needs are assessed and addressed in the round.

There is no single set of assessments universally used by learning disability teams in the UK but a number of useful tools are available. There is some guidance available (NICE/SCIE 2006; Burt and Aylward, 2000). The tests used should include some direct assessment with the person with Down Syndrome and some questionnaire assessments with a well-informed carer.

When concern arises about possible deterioration, a multi-disciplinary and multi-agency response is required. It is essential to embed the baseline screening within a Care Pathway that is part of the intellectual disability service's general approach to referrals for possible dementia. A typical pathway commences with general health screening for the reasons referred to above. Many carers and indeed GPs are not aware, for example, of the need for routine thyroid function tests for adults with Down Syndrome. There is a useful handout from the Down's Syndrome Association (Dennis, 2000) available freely on the internet that can be used to raise awareness of this issue. If the results of the health screen do not explain the presenting problem/s then a dementia assessment should be carried out with the final differential diagnosis falling to the consultant psychiatrist in intellectual disability.

If, in due course, a diagnosis of probable DAT or other type of dementia is made, careful attention will be needed to many aspects of the person's health and social care. The life expectancy after diagnosis in this group is variable and there is a lack of up-to-date information regarding the impact of new treatments. For those with the diagnosis, medication should be prescribed in line with the NICE recommendations and referrals made to other members of the multi-disciplinary team as required. Each person with probable DAT needs regular review and re-assessment as their health and social care needs change with the progression of the disease. Services need to set up mechanisms to ensure a proactive and timely response before a crisis is reached. End of life issues represent a particularly sensitive area.

What works - accommodation

Best practice is to facilitate ageing and caring in place (Janicki et al, 2002) for those people with intellectual disability who develop dementia while they are living in a supported residential setting. Moving people from a home where they are settled because the staff, for whatever reason, cannot cope with their deterioration may be detrimental to the person's well-being, particularly at a time when they are increasingly confused (Kerr, 1997). Moving people can not only be detrimental to the individual being moved and possibly to their close friends in the same home, but also

entails enormous efforts on the part of health and social care staff. Sometimes people are moved several times in quick succession and some are moved many miles away because local provision cannot be found.

Ensuring that homes can provide caring in place is complex, requiring attention to aspects as varied as the physical environment of the home, an organisational culture reflected in coherent policies and a commitment of resources, staff training and knowledge (Janicki et al, 2002; Wilkinson et al, 2004).

What works - training and awareness raising

The presence of people with Down Syndrome suffering from dementia in residential settings is increasing. Care staff need to be aware of the risks, the early signs and good practice in caring for people with dementia. There is an increasing array of useful resources available. For example a resource pack produced by BILD (Dodd et al, 2002) for carers and support staff is an invaluable toolkit to inform staff training and for homes to purchase for their own use. The Down's Syndrome Association has produced a workbook for staff (Dodd et al, 2006) that is designed for a keyworker to complete on behalf of a named person with Down Syndrome in any form of supported or residential care. The book gives guidance on offering the most effective care possible as the person's condition progresses and their needs change.

There are useful booklets for service users published by BILD (Dodd et al, 2005a; 2005b; 2005c). Service users living alongside a peer who is becoming confused need explanations and support. Lyngaard and Alexander (2004) reported on a short course for service users living with someone with dementia.

What approaches look promising?

The NICE SCIE Guideline (2006) sets a context that is, for the first time, inclusive of people with intellectual disability alongside the mainstream population. It would be reasonable to expect that this inclusiveness will be taken up in other parts of the UK currently not covered by NICE. Guidelines for good practice for supporting people with a learning disability and dementia to stay in their own homes and communities are due to be published following an ethnographic and action research project by Forbat and colleagues at the University of Edinburgh (Aug 2003-July 2007; see Forbat, 2004). The Royal College of Psychiatrists and the British Psychological Society are due to publish good practice guidelines for psychiatrists and psychologists in this area in 2008.

Currently there is a plethora of commissioning arrangements across different parts of the UK. The trend is towards integration of services with pooled budgets and increasing use of Direct Payments and Individual Budgets. This provides opportunities to improve services and avoid inter-agency buck-passing that can for example result in relatively young people with Down Syndrome and DAT being inappropriately placed in old people's homes. A successful approach requires strong local advocates and might be best achieved where service users and their carers can form a coalition with providers and managers of services.

What we need to find out

Since the NICE guideline is recent, its impact, particularly on people with Down Syndrome, has yet to be assessed. There is currently no gold standard tool for screening or diagnosis, so clearly more research is needed. There is little research on what determines a residential home's capacity to cope with clients with dementia. Few attempts have been made to capture the views of residents who themselves have dementia. There is uncertainty amongst professionals about how to cope with end-of-life issues in this population. These are all areas where the evidence base needs to be strengthened.

Written for **research in practice for adults** by Judith McBrien, Consultant Clinical Psychologist, Plymouth PCT and Visiting Professor, University of Plymouth, and Stephen Brown, Consultant Psychiatrist, Cornwall Partnership Trust, and Honorary Professor, Peninsula Medical School.

References

- Burt DB and Aylward EH (2000) Test for the diagnosis of dementia in individuals with intellectual disability, *Journal of Intellectual Disability Research*, 44, 175–180
- Dennis J (2000) *Thyroid disorder among people with Down's Syndrome. Notes for parents and carers*, London: Down's Syndrome Association
- Department of Health (2001) *Valuing People: A New Strategy for Learning Disability for the 21st Century*, London: The Stationery Office
- Dodd K, Kerr D and Fern S (2006) *Down's Syndrome and Dementia Workbook for Staff*, London: Down's Syndrome Association
- Dodd K, Turk V and Christmas M (2002) *Down's Syndrome and Dementia Resource Pack*, Kidderminster: BILD
- Dodd K, Turk V and Christmas M (2005a) *About Dementia: for people with learning disabilities*, Kidderminster: BILD
- Dodd K, Turk V and Christmas M (2005b) *About My Friend: for friends of people with Down's syndrome and dementia*, Kidderminster: BILD
- Dodd K, Turk V and Christmas M (2005c) *The Journey of Life: how people change from babies to older people*, Kidderminster: BILD
- DSA (2007) Down's Syndrome Association www.downs-syndrome.org.uk, accessed October 2007
- Forbat L, Kerr D, Wilkinson H and CARE (2004) *Ageing with a learning disability: A critical analysis of policy*, Washington: Gerontological Society of America
- Holland AJ (1999) Down's syndrome, in MP Janicki and AJ Dalton (eds) *Dementia, aging and intellectual disabilities: A handbook*, Philadelphia: Brunner/Mazel
- Holland AJ, Hon J, Huppert FA, Stevens F and Watson P (1998) Population-based study of the prevalence and presentation of dementia in adults with Down Syndrome, *British Journal of Psychiatry*, 172, 493-498
- Janicki MP and Dalton A (2000) Prevalence of dementia and impact on intellectual disability services, *Mental Retardation*, 38, 276-288

- Janicki MP, McCallion P and Dalton A (2002) Dementia-related care decision-making in group homes for persons with intellectual disabilities, *Journal of Gerontological Social Work*, 38, 179-196
- Kerr D (1997) *Down's Syndrome and Dementia: Practitioner's Guide*, Venture Press
- Lyngaard H and Alexander N (2004) 'Why are my friends changing?' Exploring dementia in people with learning disabilities, *British Journal of Learning Disabilities*, 32, 30-34
- McBrien JA, Whitwham S, Olverman K and Masters S (2005) Screening adults with Down's Syndrome for early signs of Alzheimer's disease, *Tizard Learning Disability Review*, 10 (4), 23-32
- Minns RA (1997) Pre and perinatal conditions contributing to mental retardation, *Current Opinion in Psychiatry*, 10, 354-59
- NICE /SCIE (2006) *Dementia: supporting people with dementia and their carers in health and social care*, National Clinical guideline No 42
- Turk V, Dodd K and Christmas M (2001) *Down's syndrome and dementia: briefing for commissioners*, London: Mental Health Foundation
- Wilkinson H, Kerr D, Cunningham C and Rae C (2004) *Home for good? Preparing to support people with a learning disability in a residential setting when they develop dementia*, York: Joseph Rowntree Foundation / Pavilion Publishing. Available as a summary on <http://www.jrf.org.uk/knowledge/findings/socialcare/614.asp>

Useful links and resources

- British Institute of Learning Disabilities www.bild.org.uk
- Down's Syndrome Association www.downs-syndrome.org.uk
- NICE Alzheimer's - donepezil, galantamine, rivastigmine (review) and memantine Nov 2006 <http://guidance.nice.org.uk/TA111>
- Forbat et al research website. <http://www.crf.ac.uk/Research/care.htm>

To find out how to become a Partner in research in practice *for adults* please contact Alison Petch email: alison@ripfa.org.uk tel: 01803 869754 or write to us at Blacklers, Park Road, Dartington, Totnes, Devon, TQ9 6EQ

www.ripfa.org.uk

research in practice *for adults* is a department of Dartington - an international centre for the generation and application of new ideas.

Dartington