



# Down syndrome and dementia

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*It is good news that people with Down syndrome, like the rest of the population, are now living longer. This longevity is the result of advances in medicine, improved social conditions and changes in public attitude.*

However, as people with Down syndrome live into their sixties and seventies, this increased life span, as with every one else, brings with it the illnesses and conditions of older age. One of these is dementia.

## Frequency of dementia in people with Down syndrome

For people with Down syndrome there is an increased susceptibility to the early onset of Alzheimer's-type dementia. While reported frequencies from studies vary slightly, I have provided the findings from one study in a table below, to illustrate an overall prevalence of dementia.

Between 30 and 39 years	2%
Between 40 and 49 years	9.4%
Between 50 and 59 years	36.1%
Between 60 and 69 years	54.5%

(Prasher 1995)

Studies have also found different ages of onset of the condition varying from 51.7 years (Prasher and Krishnana 1993) to 54.2 years (Lai and Williams 1989). The Prasher and Krishnan study even found a difference between the average age of onset amongst men and women. Men had a average age of onset of 54.2 years whilst women had 49.8 years.

These findings are consistent with the age related risk of dementia in the general population but in people with Down syndrome onset appears to occur between 30 and 40 years earlier.

The figures also show us that people with Down syndrome have a much higher rate of Alzheimer's type dementia than the general population. What they also show, however, is that not every one with Down syndrome develops the condition.

## A possible link between dementia and Down syndrome

Currently, the most favoured explanation of the link between Down syndrome and dementia is the existence of a third copy of Chromosome 21 (people without Down syndrome have two copies of Chromosome 21). This chromosome is involved in the production of the beta-amyloid protein, which is found in the knots and tangles in the brains of people with Alzheimer's dementia.

Almost everyone with Down syndrome will have these knots and tangles deposited on their brain by the time they are 40. What is important to note, however, is that few of them will, at this stage, have the clinical symptoms of dementia and indeed some may never develop the symptoms. No one has yet been able to explain this fully.

## Early signs of dementia

It can be difficult to determine the early changes that occur as a result of dementia. Many of the indicators of dementia are an exaggeration of already existing behaviours and deficits that exist because of a person's learning disability. Early signs of dementia among people with Down syndrome may well be a further deterioration of an already present deficit, whereas in the general population the loss will be from an area of full competence, and so it may be more easily recognised.

Below is a list of early signs that should alert carers to the possibility of the onset of dementia in people with a learning disability.

<b>Table 2</b> Early signs of dementia in people with a learning disability
Deterioration in the ability to accomplish skills of daily living
Deterioration in short term memory
Increased apathy and increased inactivity
Loss of amenability and sociability
Loss of interest in favoured hobbies
Withdrawal of spontaneous communication
Reduction in communication skills
Disorientation and confusion
Changes in depth perception
Changes in night time sleep patterns
Increased problems with comprehension
Increased wandering
(Kerr 2007)

It is important to emphasise that these difficulties and behaviours are changes from a person's normal behaviour. If someone exhibits some of these changes then a referral should be made for an assessment to determine if the changes are an indication of dementia. The possibility that they are the result of other conditions such as urinary tract infection, chest infection, hypothyroidism, visual or hearing impairment, constipation, medication, pain etc also needs to be eliminated.

Often these early signs are missed and people are not referred for assessment and diagnosis until there is a more dramatic change. A consequence of this delay can be that the person's behaviour is misunderstood and seen as "stubborn", "challenging" and "difficult" because staff and carers misinterpret the meaning and motivation for their behaviour. Staff and carers may also continue to respond to the person as they have always done, and this can cause the person with dementia unnecessary stress, confusion and anxiety, which in turn can cause challenging behaviour.

It has been recommended that everyone with Down syndrome from the age of 30 should have a baseline assessment carried out. This assessment should be repeated every five years initially, then every three years and then annually. (Oliver 1998) This will enable a quicker and well-informed comparison of changes in skills and abilities.

It is very important to get an early diagnosis so that people are given the right support as quickly as possible.

Systems need to be in place and staff and carers need the right training and information and environments adapted.

A research project Home for Good? (Wilkinson et al 2004) looked at what happens when people with a learning disability living in residential settings develop dementia. The findings were equally relevant to people, living in supported housing and with family. The project looked at three ways of providing care and support. Here is what they are:

**Option A – Ageing in Place**

Every possible effort is made to enable the person to continue to live (and die) in their own home. The service makes constant adaptations to the home of the person to meet their changing needs. This involves training of staff in dementia care, making adaptations to the built environment and working with the people who lived with the person. (Kerr et al 2002) There is also increased use of and development of relationships with other professionals such as speech and language therapists, incontinence advisors and palliative care nurses who are used in the support package.

It is probably safe to say that most people involved in supporting people with a learning disability and dementia would choose the "Ageing in place" option. However, for a variety of reasons it is not always possible for people to remain in their home.

**Option B – In Place Progression**

In this option, the learning disability service provides a specialist resource for people with dementia. In its most pure form, this service would be a distinct and separate building and resourced with all the environmental requirements for supporting someone with dementia. The staff have a high level of expertise and knowledge about both dementia and learning disability.

**Option C – Referral Out**

This involves the person being removed from the learning disability service into older people's services, which means a move to a nursing care home for older people.

A frequent reason for people moving out of their supported, residential or parents' home is because they actually have a need for nursing care. It is undoubtedly true that people will sometimes require to be moved to hospital or nursing home care in order to receive the level of nursing care they require.

There are nursing care homes that provide excellent medical and social care and where the person with a learning disability and dementia will experience an appropriate person-centred service. This cannot, however, be assumed to be the norm and care managers,

doctors and others responsible for making the decision to move someone must be aware of the nature of the service being provided.

There are a number of reasons why the 'referral out model' should be used with extreme caution.

Firstly, staff in older people's services will often express anxiety about caring for people with a learning disability and will, often through fear and lack of knowledge and experience, fail to give adequate care. (Wilkinson et al 2004, Thompson and Wright 2001) When a decision has been made to move someone it is essential that there are clear benefits to the person. To move someone into a nursing home because there are nursing staff available but where the person may not receive adequate care in other areas such as eating, washing and skin care (Wilkinson et al 2004) is questionable. The balance of gains and losses needs to be given considered, multidisciplinary and well informed consideration.

Secondly, placing people with a learning disability, who may be in their forties and fifties if they have Down syndrome, with older people without a disability who may be in their late seventies and eighties is inappropriate in terms of meeting individual assessed needs. People with a learning disability will often have had different life experiences to the general population and these need to be reflected in the activities and interactions that occur in the home.

Thirdly, it can be the case that people are moved because they require a level of nursing care that often could and should be provided by the district nurse, the palliative care nurse and/or the staff themselves with advice and support from a nurse.

Having dementia does not mean that people suddenly lose their skills, or forget what they want and need. What is important is that systems are put in place that help people maintain their skills and continue to have positive experiences. There is a great danger that if we do not act urgently to set things up then many of the gains that have been made for people with Down syndrome will be lost when they develop dementia.

## References

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- See page 17 for information about Diana Kerr's recent Australian lecture tour and the launch of Down syndrome and Alzheimer's disease.