

Mental health and behavioural characteristics of dementia in people with Down Syndrome

People with Down Syndrome are now living longer and healthier lives. This in part is attributable to better health care and nutrition in infancy and childhood. The average life expectancy has increased dramatically over the last 60 years with many people living into their fifties and sixties. As in the general population, increased life expectancy brings with it, the risk of developing diseases of old age. One such disease is Alzheimer's disease (AD), an illness characterised by memory loss and behavioural change. Alzheimer's disease is a complex disorder that affects the brain initially with short term memory loss but also affecting skills such as speech, walking, knowledge, judgement and ultimately leading the individual to complete dependence on their carers for all activities of daily living.

The exact cause of AD is unknown, but scientists have discovered an excess of two proteins Tau and Amyloid that accumulate within brain tissue and surrounding tissue leading to death of brain cells (neurones), particularly in areas responsible for speech and memory.

For most people in the general population Alzheimer's disease is age-related, with the incidence increasing with advanced age. In some rare cases there are genetic abnormalities leading to family risk. In people with Down Syndrome there are three copies of the gene coding for the amyloid protein and simply more amyloid deposition at a younger age within the brain, with the result that about 15–40% of people with Down Syndrome develop AD at a younger age (usually mid-fifties). Research is currently investigating what factors protect people with Down Syndrome from Alzheimer's disease.

Clinical features

Usually, as in the general population, the first clinical feature of dementia is memory loss, however, this may be hard to detect.

Memory loss is due to impairment in the area of the Hippocampus, a small midline structure within the brain. The critical defect is one of loss of a neurotransmitter called acetylcholine leading to faulty connections between neurones and initially loss of newly acquired memories. Short-term memory loss may present as failure to remember names in the workshop or neighbourhood, forgetfulness of keys or personal objects, and forgetfulness about time of day. One common feature I have observed is a person forgetting that they have eaten a meal and asking for dinner at 2 am in the morning or other inappropriate times.

Other features are **impairment of speech and gait**, particularly difficulty with falling or high stepping when moving from one room to another. This is because **difficulties with perception** may make distance and height hard to judge. Some people may lose their **initiative** and need prompting to do a task, but once started they may complete that task successfully. Complex tasks that require a person to sequence the task in 2-3 steps (sorting and collating tasks in the workshop) may become difficult and frustrating.

As the illness progresses, people may develop **incontinence**, and may need assistance in **walking and feeding**. These features are not early features and the illness should have been recognised before then.

In a small proportion of people there may be **psychiatric features**, some people may appear **depressed** or some people may misinterpret their environment due to altered perception, or experience **hallucinations**. Such symptoms are due to an abnormal balance of neurochemicals such as noradrenaline, serotonin and dopamine within the brain. It is also important not to misdiagnose

dementia—both depression and psychosis may mimic dementia; sometimes adequate treatment of psychiatric illness will differentiate the correct illness. A severe depressive illness will usually be accompanied by weight loss, anorexia, early morning waking and low mood that is characteristically worse in the morning. Whilst sleep disturbance occurs in dementia, it is usually characterised by difficulty falling and staying asleep.

Epilepsy, either in the form of a generalised seizure or in a localised form (myoclonus), is very common but is not a feature associated with early dementia. It responds well to treatment and is rarely a problem once stabilised.

One mystery in people with Down Syndrome is that **several other conditions** may mimic dementia. I have mentioned psychiatric illness, but any physical illness—untreated hypothyroidism, a deficiency of vitamin B12 or folate, high levels of calcium in the blood, sensory loss (particularly visual or hearing loss), bereavement or abuse—may lead to regression in skills as discussed above. Therefore a proper assessment for dementia requires a comprehensive evaluation to exclude untreated medical or social distress, remembering that a person can have more than one illness at a time and if memory loss and skill loss do not improve with, say, grief therapy, and antidepressants have failed to treat a post-bereavement depression, this may indicate that dementia should be considered.

A comprehensive **assessment** will consist of a multidisciplinary approach with an assessment of memory and thought, physical health and mental health and social wellbeing. Occasionally when a diagnosis is complicated extra tests such as brain scans (CAT scans or MRI scans) or EEG testing may be required. At the centre where I work, if we feel that the diagnosis is unclear because of other factors we review the person at 6-monthly intervals. Since dementia is a progressive illness, we repeat the assessment to see if decline (and therefore dementia) is present.

If dementia is present, then the **management** is multidisciplinary and aimed at providing a person-centred approach aiming to preserve independence for as long as possible. Specific management of mental health problems may involve the use of drugs that increase the amount of acetylcholine in the brain (e.g. donepezil, galantamine and rivastigmine) to improve memory. Memantine may be used in middle-stage dementia to stabilise memory. Where depression is a problem, selective serotonin reuptake inhibitors (SSRIs) may be helpful. Hallucinations and delusions may be helped by dopamine inhibitors, but these are used cautiously and at low dose because of possible side effects with older drugs in this class and because some of the newer drugs, whilst having fewer side effects, have been implicated in multi-infarct dementia.

Sleep disturbance is best treated environmentally but if it is still a problem it may be helped by trazadone which acts to increase serotonin. It is best to avoid sedatives in dementia as they may add to confusion and if hypnotics are used they should be used for a short period only and at the lowest dose.

To summarise, Alzheimer's disease occurs with increased incidence in people with Down Syndrome, usually after age 50. Memory loss and impairments of thought and behaviour are key features and it is important to differentiate dementia from other illnesses. Whilst there is as yet no cure for AD, many symptoms may be helped by environmental, behavioural and medical intervention.

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