

What is Alzheimer's disease?

Dementia is a syndrome consisting of a number of symptoms that include a reduced ability to perform familiar tasks, impairment of memory, judgment and reasoning, and changes in mood and behaviour. Some dementias are caused by treatable conditions such as depression, thyroid disease, infections or drug interactions. However, treatments are not yet available for the progressive, irreversible, dementias in which nerve cells in the brain become sick and eventually die.

According to a 2016 study commissioned by the Alzheimer Society of Canada, the number of Canadians living with dementia now stands at 564,000¹. Other forms of dementia resemble Alzheimer's disease in that they also involve a progressive degeneration of brain cells that is currently irreversible. They include the dementia associated with vascular dementia, Lewy body dementia, frontotemporal dementia, Creutzfeldt-Jakob disease and mixed dementia. The brain abnormalities that occur with Alzheimer's disease can start as early as one's 30's or 40's, however, the gradual onset of dementia symptoms usually begins in one's 60's or 70's. Until then, the brain's self-repair mechanisms seem able to compensate for the ever-increasing nerve cell sickness. Improved testing techniques, plus the willingness of people to consult a doctor at the first signs of behavioural and memory impairment, are now leading to earlier diagnoses of Alzheimer's disease.

What is Down syndrome?

Down syndrome (DS) is a genetic disorder in which an individual has three copies of the 21st chromosome instead of two. In Canada, the incidence of Down syndrome is approximately 1 in every 750 live births². The condition is not related to gender, race, nationality or socio-economic status and the exact cause of Down syndrome is still not known.

Down syndrome is the most common genetic cause of severe learning disabilities in children³ associated with developmental delays, learning difficulties, health issues, and some physical abnormalities. Individuals with Down syndrome vary in their abilities, and it is important to recognize that each person has unique attributes and strengths.

Life expectancy of individuals with Down syndrome has increased due to improvements in health care and decreased infant mortality. The life expectancy of an individual with Down syndrome tends to be in the 60's. Persons with Down syndrome may be predisposed to certain illnesses and medical conditions, but that genetic arrangement does not guarantee their development. The same illnesses and conditions are also present in the general population⁴.

1 Alzheimer Society of Canada (2016).

2 Congenital Anomalies in Canada: A Perinatal Surveillance Health Report, Public Health Agency of Health Canada (2013).

3 MayoClinic.org (2014).

4 The Canadian Down Syndrome Society (2009).

What are some of the warning signs of dementia in a person that has Down syndrome?

The first sign of dementia in individuals with Down syndrome is often changes in their behaviour and personality (Ball et. al, 2006), which differ somewhat from the warning signs of Alzheimer's disease especially in relation to memory impairments and language. In an individual with Down syndrome, it is essential to pay attention to behavioural changes such as reduced empathy, social withdrawal, emotional instability and apathy (Ball et. al, 2006). Researchers have discovered that these changes often progress to more closely resemble characteristics of frontal temporal dementia (Ball et. al, 2006). Individuals with Down syndrome and a diagnosis of frontal temporal dementia are 1.5 times more likely to develop Alzheimer's disease (Ball et. al, 2006). Another significant warning sign of dementia is if an individual with Down syndrome is experiencing seizures for the first time in their life (British Psychological Society, 2015).

How is Down syndrome associated with Alzheimer's disease⁵?

The hallmarks of Alzheimer's disease are the presence of "plaques" and "tangles" in the brain. The major constituent of the plaques is a protein ("A-beta") which is split off from a much larger parent protein called Amyloid Precursor Protein (APP). The tangles are inside the affected nerve cells, and they may be induced to develop by the accumulating A-beta outside the cells. Between them the A-beta and the tangles cause the brain cells to become sick, and eventually to die. The sickness begins in certain portions of the brain and then tends to spread. This explains why various abilities are altered during the progression of the illness.

Researchers have found that the production of APP is linked to chromosome 21. Since individuals with Down syndrome have an additional 21st chromosome, they are prone to an over-production of APP. As indicated above, more APP is likely to lead to more A-beta production, and most individuals with Down syndrome will indeed develop the plaques and tangles in the brain characteristic of Alzheimer's disease. Nevertheless, not all of them go on to exhibit symptoms of the disease⁶. It is thought that other factors may impact on the development of the disease such as: family history of Alzheimer's disease, premature aging, environmental factors and the role of unknown genes.

The prevalence rates of Alzheimer's disease in adults with Down syndrome vary according to age. 25% of individuals with Down syndrome over the age of 35 display the symptoms commonly associated with Alzheimer's disease (NDSS, 2009), and overall the incidence of Alzheimer's disease in the Down syndrome population is estimated to be three to five times greater than the general population (NDSS, 2009). As with the general population, the chances of those with Down syndrome developing the plaques and tangles characteristic of Alzheimer's disease increase with age.

Baseline Screening and Monitoring⁷

Ideally, at age 30 every individual with Down syndrome should have testing done to assess their cognitive and functional abilities. This baseline can be used when assessing if the individual has experienced a decline in cognitive ability or functioning. It can be helpful to conduct regular testing to monitor cardiovascular health, diabetes and to differentiate between other conditions that may present similar symptoms as dementia.

5 Prasher, V., *Alzheimer and Dementia in Down Syndrome and intellectual disabilities*. Radcliffe Publishing (2005).

6 Alzheimer Society of the UK (2010).

7 *Dementia and People with Intellectual Disabilities*, The British Psychological Society, April 2015

Some of these useful tests include:

- Yearly testing of thyroid function
- Yearly testing of fasting blood glucose and lipids (including cholesterol and triglycerides)
- Eye exams every year
- Hearing tests every two years
- B12 and Folate level
- Urea and electrolytes
- Liver function tests

How is a person with Down syndrome assessed for dementia?

As in diagnosing anyone with possible dementia, it is vital to rule out any other physical conditions or other possible explanations. Some of the changes may be caused by depression, thyroid problems, trauma or abuse, sensory issues, infections, or other life events (Dodd, 2009). The challenge in diagnosing individuals with Down syndrome is that many of the diagnostic tools used for the general population may not be suitable. These tools do not consider the intellectual skills and status of individuals with Down syndrome and may present an inaccurate representation of the situation. An individual's doctor should perform tests tailored specifically for individuals with Down syndrome.

Some of these tests are:

- The Dementia Scale for Down syndrome (DSDS)
- The Test for Severe Impairment (TSI)
- The Down Syndrome Mental State Exam (DSMSE).
- The Dementia Questionnaire for People with Learning Disabilities (DLD)

Regardless of what assessment tools are used, it remains vital that family members and health care providers document any changes in behaviour, routine and mood. These observations are valuable in assisting the doctor in assessment and diagnosis. It is also important to ask the affected person to describe the changes that they are experiencing (DSA, 2004). The person's account and the family's observations coupled with a physical examination will assist in the process of making a diagnosis. It is important to note that none of these tests can conclusively determine if an individual with Down syndrome has dementia. These tests coupled with feedback from caregivers provide the basis for a probable diagnosis of an individual.

What does the progression of the disease look like for an individual with Down syndrome?⁸

The progression of Alzheimer's disease in individuals with Down syndrome follows similar stages as anyone with Alzheimer's disease would experience⁹. However, it is important to note the differences in the progression of the disease for individuals with Down syndrome.

Depression:

Individuals with Down syndrome often have more depressive symptoms related to their dementia.

Seizures:

Additionally, an adult with Down syndrome and Alzheimer's disease is increasingly vulnerable to falls. This can be due to a possible history of seizures, cognitive impairment, and possible sleep medication usage. In the late-stage of Alzheimer's disease (among people who do not have DS), 15-25% of individuals can experience seizures. However, in individuals with Down syndrome and Alzheimer's disease, 90% will experience seizures. The presence of seizures is often the first indicator of dementia in an individual with Down syndrome (Prasher, 2005). Therefore, it is necessary to seek further medical advice if an individual with Down syndrome is experiencing seizures.

Is there Treatment?

The majority of studies about drug treatment options for Alzheimer's disease have focused on the use of the drugs with individuals who do not have Down syndrome. Most of the studies that have concentrated on the use of the drug within the Down syndrome population have been related to Donepezil (also known as Aricept).

There is a lack of investigation into the impact of other drugs (Rivastigmine, Galantamine, Memantine) with individuals who have Down syndrome (Prasher, 2005). Research studies tell us that we need to consider common health concerns associated with Down syndrome, such as thyroid conditions and diabetes. Individuals with Down syndrome tend to show symptoms of Alzheimer's disease at a much younger age than the rest of the population (Cochrane Library, 2009). As well, their body size, metabolism and heart rate may influence the way that common Alzheimer's disease drugs are prescribed in this population (Cochrane Library, 2009). The Alzheimer Society of Canada publishes information on the medications commonly prescribed for individuals with dementia. These sheets describe how the medication can assist individuals, outline dosage information and explain things to consider before taking each particular drug. This information can be a useful resource when evaluating medication options for someone with Down syndrome and Alzheimer's disease.

⁸ Prasher, V., *Alzheimer and Dementia in Down Syndrome and intellectual disabilities*. Radcliffe Publishing (2005).

⁹ The Alzheimer Society of Canada developed the Progression series, a five-part series on the stages of Alzheimer's disease, which is written for the person with the disease, their family and caregivers. The Progression series are available at: <http://www.alzheimer.ca/stages>

The Edinburgh Principles¹⁰

The following principles were developed by international researchers and organizations that provide services to individuals with Down syndrome and Alzheimer's disease. When caring and interacting with these individuals it is recommended that families, caregivers and health care professionals adhere to these principles. These principles are transferable to individuals with dementia that do not have Down syndrome, and so can help guide your interactions generally with people living with dementia.

1. Adopt an operational philosophy that promotes the utmost quality of life of persons with intellectual disabilities affected by dementia and, whenever possible, base services and support practices on a person-centred approach.
2. Affirm that individual strengths, capabilities, skills, and wishes are overriding considerations in any decision-making for and by persons with intellectual disabilities affected by dementia.
3. Involve the individual, her or his family, and other close supports in all phases of assessment and services planning and provision for the person with an intellectual disability affected with dementia.
4. Ensure that appropriate diagnostic, assessment and intervention services and resources are available to meet the individual needs, and support healthy aging, of persons with intellectual disabilities associated with dementia.
5. Plan and provide supports and services that optimize the potential of adults with intellectual disabilities affected by dementia to remain in the chosen home and community.
6. Ensure that persons with intellectual disabilities affected by dementia have the same access to appropriate services and supports as afforded to other persons in the general population affected by dementia.
7. Ensure that generic, cooperative, and proactive strategic planning across relevant policy, provider and advocacy groups involves consideration of the current and future needs of adults with intellectual disabilities associated with dementia.

Providing Support¹¹

The following suggestions are useful for all individuals with dementia and can be adopted for use by individuals with Down syndrome and Alzheimer's disease.

- Help create as many opportunities as possible for an individual to make choices and have control in their life.
- Create a routine to provide structure and security for the individual.
- Visual cues such as labels on doors can make it easier for some individuals to find their way around their home.
- Use body language to assist in communication.
- Incorporate massage and aroma therapy to promote relaxation.
- Examine the individual's environment and make modifications if necessary. Sometimes if the environment is too noisy or distracting this can increase agitation levels in an individual with dementia. It is important to minimize these disturbances to help calm the individual.
- Use stories or scrapbooks to reminisce about the individual's life and interests.

¹⁰ Wilkinson, H. et al., The Edinburgh Principles with Accompanying Guidelines and Recommendations (2001). This document can be found at <https://www.iassidd.org/uploads/legacy/pdf/edinburg-principles.pdf>

¹¹ Alzheimer Society of the UK (2010).

Additional suggestions¹²:

- Encourage ongoing dialogue with the person to determine if they understand the changes that are happening. Check in to see how these changes are making them feel.
- Help them to remain a part of their social network and to maintain friendships.
- Consider current abilities and adapt routines accordingly.

Special Considerations for Daily Living

The Alzheimer Society has published information on modifying daily activities for people with dementia according to their current abilities. The information below has been identified by researchers as issues that are specific to individuals with Down syndrome who have dementia.

Hearing¹³:

The progression of dementia makes it increasingly difficult for someone to express themselves and/or to understand what is being said to them by their caregivers. This situation coupled with hearing loss can make it increasingly challenging to communicate with someone who has Down syndrome. It is common for individuals with Down syndrome to have problems with their hearing. As individuals age, they may experience further hearing loss and have trouble distinguishing different frequencies and/or tones. Regular assessments should be undertaken to determine the cause of the hearing loss, to address excess earwax and treat possible infections. Individuals with hearing aids may need assistance or cuing to use them as the disease progresses. Strategies to improve communication include using visual aids, minimizing distractions, decreasing background noises, and ensuring adequate lighting. Make sure to face the individual when speaking to enhance communication.

Dental Care¹⁴:

Good oral care is essential to prevent the loss of teeth. Chewing on the back teeth actually helps to decrease the build up of ear wax in an individual with Down syndrome. As the disease progresses it may be difficult for individuals to communicate if they are experiencing pain due to dental issues. This may delay treatment and cause eating difficulties for the individual with dementia.

Mobility¹⁵:

Individuals with Down syndrome may have problems finding proper fitting shoes because many people have shorter and wider feet. This can exacerbate the challenges that may occur with mobility as the disease progresses. Another issue is the need to adapt seating because individuals with Down syndrome typically have a shorter stature. If seating is not adapted individuals may be at a greater risk of accidents when moving around. Inner ear infections can increase the likelihood of mobility problems. It is recommended that health care professionals such as occupational therapists, podiatrists, or physiotherapists be actively involved to minimize the risk of injury for the individual.

12 Dodd, K., et al., *Down's syndrome and dementia*. BILD (2009).

13 Ibid.

14 Ibid.

15 Ibid.

Pain¹⁶:

In people with Alzheimer's disease and a learning difficulty, pain is often under-treated. With someone who has dementia, the ability to communicate can become challenging as the disease progresses. When someone has Down syndrome they may have less vocabulary to begin with, so the loss of additional words further compromises the person's communication with others. If the individual has a history of certain behaviours caregivers and health care professionals may be less likely to recognize that the individual is in pain. Also, when a person with Down syndrome and Alzheimer's disease has difficulty sleeping this may not be a result of the dementia but a problem with pain. Another issue is the role that medication such as anti-psychotics can play in concealing pain. Researchers in this field emphasize the importance of learning as much as possible about the person's history and their communication style. This information can be useful in assisting caregivers and staff to identify communication problems or recognize the presence of pain. Common causes of pain in individuals with Alzheimer's disease and Down syndrome are dental problems, arthritis, impacted earwax, eye infections, urinary tract infections and constipation.

Planning Ahead¹⁷

The progressive nature of dementia necessitates that individuals and their family plan ahead for the changes that will occur. This planning includes outlining preferences for the care that they receive during their life and their wishes for end of life care.

An important part of this planning is getting to know the person with Down syndrome. This includes learning about their interests, ways of communicating and things that bring them joy/comfort. This knowledge becomes increasingly useful when the progression of the illness makes it challenging for the individual to communicate. Caregivers can use this information to plan outings or activities more suited to the individual's personal preferences. Having this knowledge allows caregivers to understand what may be causing agitation or soothe an individual when they become upset.

It is essential to find out what the person knows about the illness and the different care options available. Knowing the latter information can assist the caregiver or health care professional to plan the subjects to include in the conversation. This can be a difficult discussion to have but the individual should gain some reassurance by knowing that their care will be guided by their preferences and wishes.

For more information:

Please contact your local Alzheimer Society for information and support. Visit our website at www.alzheimer.ca.

More information can be obtained from the following organizations:

1. The Canadian Down Syndrome Society: www.cdss.ca
2. Down syndrome Research Foundation: www.dsrf.org

¹⁶ Kerr, D., et al., *Responding to the pain experiences of people with a learning difficulty and dementia*. Joseph Rowntree Foundation (2006).

¹⁷ Leigh Ann Creaney Kingsbury, *Planning Ahead: a Guide to Communicating Health Care and End of Life Wishes*. American Association on Intellectual & developmental Disabilities (2009).

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