Alzheimer Society

Down syndrome and Alzheimer's disease

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 damaged and eventually die. Alzheimer's disease is the most common type of dementia, and currently there is no cure available.

In 2022, the Alzheimer Society of Canada released the Landmark Study Report, stating that there are 600,000 people in Canada living with dementia[1]. Other types of dementia resemble Alzheimer's disease in that they also involve a progressive degeneration of brain cells that is currently irreversible. Some other types of dementia are 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, for most people symptoms usually begin in one's 60's or 70's. Improved testing techniques, plus the willingness of people to consult a doctor at the first signs of symptoms, are now leading to earlier diagnoses of Alzheimer's disease.

What is Down syndrome?

Down syndrome 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 births[2]. 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 children[3] 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 population[4].

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1 Alzheimer Society of Canada (2022). 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). This differs 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 frontotemporal dementia (Ball et. al, 2006). Individuals with Down syndrome and a diagnosis of frontotemporal 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 disease[5]?

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 damaged, and eventually to die. The damage 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 disease[6]. 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 percent of individuals with Down syndrome over the age of 35 display the symptoms commonly associated with Alzheimer's disease (NDSS, 2009). The overall 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 Monitoring[7]

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.

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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 Alzheimer's 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 experience[9]. 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, a person 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 Down syndrome), 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 a person 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. This information describes how the medication can assist individuals, outlines dosage information and explains 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. You can view this information at alzheimer.ca/medications.

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 is available at: www.alzheimer.ca/stages

The Edinburgh Principles[10]

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, their 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 support[11]

The following suggestions are useful for all people 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 person.
- Visual cues, such as labels on doors, can make it easier for some people 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 person.
- Use stories or scrapbooks to reminisce about the person's life and interests.

10 Wilkinson, H. et al., The Edinburgh Principles with Accompanying Guidelines and Recommendations (2001). 11 Alzheimer Society of the UK (2010).

Additional suggestions[12]:

- 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 created resources on how to modify 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 people with Down syndrome who have dementia.

Hearing[13]:

The progression of dementia makes it increasingly difficult for someone to express themselves and/or to understand what is being said to them by others. This situation coupled with hearing loss can make it increasingly challenging to communicate with someone who has Down syndrome. It is common for people 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. People with hearing aids may need assistance or cuing to use them as their dementia progresses. Strategies to improve communication include using visual aids, minimizing distractions, decreasing background noises, and ensuring adequate lighting. Make sure to face the person when speaking to enhance communication.

Dental care[14]:

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 dementia 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 person with dementia.

Mobility[15]:

People with Down syndrome may have problems finding proper fitting shoes because many people have shorter and wider feet. This can increase the challenges that may occur with mobility as the dementia progresses. Another issue is the need to adapt seating because people 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 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.

Pain[16]:

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 help assist 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 ahead[17]

The progressive nature of dementia means that individuals and their family should 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 and comfort. This knowledge becomes increasingly useful when the progression of dementia makes it challenging for the person 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 a person when they become upset.

It is essential to find out what the person knows about dementia 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 alzheimer.ca.

More information can be obtained from the following organizations:

1. The Canadian Down Syndrome Society: cdss.ca
2. Down Syndrome Research Foundation: 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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