

Down
syndrome

and
Alzheimer's
disease



An Australian Government Initiative



Down Syndrome
AUSTRALIA NETWORK



Alzheimer's
Australia
Living with dementia





An Australian Government Initiative

This publication was supported by funding from the Australian Government Department of Health and Ageing, under the Peak Partnership Project.

The views expressed in this work are the views of its authors and not necessarily those of the Commonwealth of Australia.

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ISBN 978-1-921570-03-2

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About Alzheimer's disease and Down syndrome

Down syndrome is the most common cause of intellectual disability and occurs across all races and cultures. Down syndrome is associated with a wide range of health issues.

This booklet will help you understand the relationship between Down syndrome and Alzheimer's disease.

Alzheimer's disease is the most common form of dementia in the general population. It is also the most common form of dementia in people with Down syndrome.

The difference for people with Down syndrome is that it occurs more frequently and at a younger age than in the general population.

What is Alzheimer's disease?

Alzheimer's disease is a progressive degenerative condition of the brain that results in a gradual change, over years, in a person's ability to think and remember and to perform tasks of daily living.

Why do people with Down syndrome often get Alzheimer's disease?

Most people with Down syndrome have three copies of chromosome 21. This is known as "trisomy 21". The remaining 5% have other variations, mosaic or translocation Down syndrome, where the pattern of chromosomes is a little different. This means that most people with Down syndrome have three copies of all genes coded on chromosome 21, while people without Down syndrome only have two copies.

A specific brain protein called amyloid precursor protein (APP) is the protein that is thought to be associated with Alzheimer's disease. The gene that codes for APP is located on chromosome 21. For more information on the technical process that occurs with APP and Alzheimer's disease visit the Alzheimer's Australia website.

Having three copies of the APP gene results in excessive production and depositing of the amyloid protein in the brain. A complex sequence of events leads to the development of plaques and tangles - the classic microscopic findings of Alzheimer's disease in the brain - and the loss of brain cells. The progressive loss of brain cells results in the symptoms of Alzheimer's disease.

Do all people with Down syndrome develop Alzheimer's disease?

No. Not all people with Down syndrome will get the dementia of Alzheimer's disease.

Studies have shown that the microscopic plaques and tangles in the brain that are associated with Alzheimer's disease are seen in almost all people with Down syndrome by the age of 40 years. However, the presence of plaques and tangles in the brain does not necessarily mean that a person will show the clinical symptoms of dementia. Research using new technologies is needed to help us understand the relationship between the changes to the brain and the extent to which a person develops symptoms of dementia.

It is most important not to assume that changes that you may notice in an older person with Down syndrome are due to dementia.

How common is Alzheimer's disease in people with Down syndrome?

Research reports different rates of Alzheimer's disease in people with Down syndrome. Most studies report about 50% of people with Down syndrome will develop Alzheimer's disease by the age of 60 years. This is an important finding as it means that about 50% of people with Down syndrome in their 50s do not have Alzheimer's disease. Recent studies also indicate that the average age of diagnosis of Alzheimer's disease in people with Down syndrome has been increasing over time from 50 years to the mid 50s. This could be due to children with Down syndrome growing up with their families, early intervention and education, better nutrition and health care and enriched adult life.

Can Alzheimer's disease be prevented?

As with the general population, little is known about whether Alzheimer's disease can be prevented in people with Down syndrome. However, the average age of diagnosis of Alzheimer's disease in people with Down syndrome has been increasing suggesting that improvements in health care, education and adult occupation may be having a positive effect on the brain.

While it is not yet known how to prevent Alzheimer's disease, adopting the Mind your Mind® lifestyle may reduce the risk of developing dementia. This involves:

- Maintaining good heart health – what is good for the heart is usually good for the brain – through healthy diet and exercise;
- Good dental care;
- Maintaining good mental health;
- Maintaining social networks and activities; and
- Keeping the brain active – with music, art, drama, sport, reading, work etc.

For more information about the Mind your Mind® program, visit the Alzheimer's Australia website.

It is recommended that the child or adult with Down syndrome should have annual health assessments. Medicare funds annual health assessments of people with intellectual disabilities by their general practitioner. Hearing and vision should be tested every 1-2 years.

Some people find it helpful to keep a folder – or personal health record – of copies of reports by health professionals and other health information. If you are interested an example of a personal health record can be found under the 'Products and Resources' section of the Centre for Developmental Disability Health Victoria website www.cddh.monash.org

Presenting this information at clinical appointments is often helpful for the clinician, especially if the clinician is not familiar with the person.

Diagnosis



How is Alzheimer's disease diagnosed in someone with Down syndrome?

The process of making a diagnosis of Alzheimer's disease in someone with Down syndrome is the same as making a diagnosis of Alzheimer's disease in anyone else. Diagnosis is based on a detailed history of progressive change over time in thinking, memory and daily living skills, physical examination, tests of thinking and memory, and investigations (blood tests and brain scans) to rule out other causes of decline in functioning.

The difference for people with Down syndrome is that they have pre-existing difficulties with thinking, remembering and daily living skills. This means that people with Down syndrome generally don't do as well on the standard tests used in the general population, making interpretation of results difficult unless there are earlier assessments for comparison. Therefore it is important for the clinician to have a very clear picture of the person's abilities before any change was noticed and how the person's abilities have progressively changed.

What information should be passed on to medical professionals to help with a diagnosis?

If the child or adult with Down syndrome has speech, psychological, educational or other assessments, it is a good idea to keep copies of any reports. If possible, it is recommended that adults with Down syndrome have a professional assessment of communication, memory and other thinking skills, preferably during their 20s.

Opportunities for formal assessment of communication, memory and thinking may be limited. However a simple, but very helpful, thing you can do is to begin collecting and dating information as early as possible (i.e. throughout adolescence and early adulthood). This information can inform clinicians about someone's abilities before there were any signs of change in abilities.

Here are some ideas.

- Collect and date examples of writing, drawings, paintings and any other art and craft.
- Keep an annual record of:
 - > The person's ability to complete chores or other responsibilities at home;
 - > Involvement in hobbies and recreational activities;
 - > Work activities of all types (paid, voluntary);
 - > Routines; and
 - > Personality and behaviour.

The more detailed the description the more useful it will be in the future.

- Take photographs and video tapes of the person involved in daily activities such as:
 - > Preparing a cup of tea or coffee or a simple meal;
 - > Putting on clothes, especially doing up buttons and shoe laces;
 - > Playing sports; and
 - > Engaging in 10-20 minutes of conversation or interaction with a family member, friend or other familiar person.

How Does Alzheimer's disease affect people with Down syndrome?

Alzheimer's disease affects people with Down syndrome in the same way as it affects other people. Changes will vary from person to person, and depends to some extent on a person's previous strengths and areas of difficulty.

Changes seen in people who have Alzheimer's disease may include difficulty in the skills listed below:

1. Executive skills

- Organising and completing complex tasks (such as getting dressed or making a cup of tea) that they previously have been able to do independently, without needing to be prompted to start or to complete each step of the task.

2. Memory

- Remembering the answer to questions asked – e.g. they may repeatedly ask the same question;
- Remembering what they did this morning or yesterday;
- Remembering that they have already done something - eg. they may do it again;
- Remembering where they have put things; and
- Remembering names of people.

Note: People with Alzheimer's disease may have difficulty remembering events that have just happened, but may remember past events very well.

3. Language skills

- Finding familiar words;
- Using complex sentences;
- Participating in conversations;
- Understanding what has been said; and
- Reading and writing.

4. Recognition/Knowing what something is

- Recognising people, objects or places.

5. Visual-spatial skills

- Telling left from right;
- Knowing where parts of the body are;
- Finding their way along streets that previously were well known;
- Finding their own room or the toilet;
- Negotiating patterned floors or moving from one floor surface to another;
- Negotiating stairs, curbs and escalators; and
- Complexity and detail in drawing or artwork.

6. Learnt motor skills

- Doing up shoe laces, zips and buttons;
- Folding clothes;
- Using cutlery and cups;
- Grooming;
- Swallowing; and
- Walking.

7. Psychological and behavioural

In addition to a decline in abilities, the person with Alzheimer's disease may have psychological and behavioural changes including:

- Loss of motivation;
- Sleep disturbance;
- Wandering;
- Agitation;
- Resistiveness;
- Irritability and aggression;
- Mood changes - e.g. depression; and
- Delusions and hallucinations.

It is important to remember that behavioural and psychological changes may be caused by other medical conditions or responses to the person's circumstances.

8. Seizures

Epilepsy occurs more often in people with Down syndrome than in the general population. There is an association between seizures and Alzheimer's disease including:

- Onset, or worsening of pre-existing epilepsy, particularly in the later stages of Alzheimer's disease; and
- Development of myoclonic jerks (little motor jerking).

My family member has shown some of these symptoms, do they have Alzheimer's disease?

A decline in abilities in an adult with Down syndrome is not always due to Alzheimer's disease. Adults with Down syndrome may have other physical or mental health problems that may cause a decline in their abilities. Treatment of these conditions can lead to great improvements in the person's functioning and well being. Medications may also cause a decline in functioning, especially medications for seizures, behaviour problems or psychiatric conditions. Pain, stress and changes in environment may also cause changes in a person's capabilities. Rapid decline is more likely to be due to a

problem other than Alzheimer's disease. If you have any concerns, see your general practitioner as soon as possible.

Some of the common causes of a decline in abilities in someone with Down syndrome that are not due to Alzheimer's disease include:

Psychiatric Disorder

- Depression;
- Grief;
- Adjustment Disorders;
- Psychosis; and
- Delirium (confusion due to medical conditions or medications).

Sensory impairments

- Hearing; and
- Vision.

Musculoskeletal Problems

- Arthritis, especially of the spine, hips and knees;
- Muscle weakness;
- Lack of physical fitness; and
- Bone fractures.

Medical conditions

- Hypothyroidism;
- Sleep apnea;
- Heart problems;
- Chest infection;
- Urine infection;
- Constipation; and
- Epilepsy.

Medications

Other Brain Disorders

- Vascular changes in brain.

If you have any concerns, see your general practitioner as soon as possible.

Where can I get advice about assessment services?

Your general practitioner can refer the person to local public or private services for assessment. You can also contact your local Down Syndrome Australia or Alzheimer's Australia office for more information.

Are there treatments for Alzheimer's disease?

The treatments for a person with Down syndrome and Alzheimer's disease are the same as for the general population. Currently there are no treatments that halt or even slow down the progression of Alzheimer's disease. However there are a few medications that may provide some improvement in cognitive function and quality of life. However, people with Down syndrome may require smaller doses and may be more likely to develop side effects. A specialist such as a psychiatrist, geriatrician or neurologist may prescribe medications that help to improve thinking in people with Alzheimer's disease. For more information on these medications, please refer to the fact sheets on the Alzheimer's Australia website.

The medications used to treat depression and other medical conditions, such as seizures or chest infections, are just as important if not more important, as the medications for Alzheimer's disease. If you have any questions regarding medications you should contact your local general practitioner or health specialist.

Support



What can I do to support a person diagnosed with Alzheimer's disease?

- Make sure that you look after yourself by having periods of rest, respite and recreation.
- Routines are really important for the person, keep these going.
- Keep the person active, but:
 - > Reduce demands made on the person;
 - > Break tasks into simpler steps ; and
 - > Support the person to do things for themselves by compensating for skills that are deteriorating or lost, such as serving finger foods, laying the person's clothing out, changing to shoes without laces.
- Check that the environment is suitable for someone with dementia:
 - > Remember the person may have problems with their 3D vision; changes in floor colour may look like changes in level;
 - > Shiny floors could look like water;
 - > People often have difficulty with the colours at the bottom end of the spectrum. Try to use red, orange and yellow for things that you want people to see e.g. toilet doors;
 - > Disguise doors you do not want the person to go through;
 - > Think about the impact of mirrors. The person may not recognise themselves in the mirror. They may think their reflection is stranger in their room; and
 - > Minimise confusion by reducing large choices, clutter, noise and glare in the lighting.
- Use strategies, such as redirection, and avoid confrontation as distress can increase confusion.
- Think about ways to explain to the person what's about to happen or is happening.
- Make use of pictures:
 - > To represent people (e.g. staff and friends at day placements);
 - > To represent planned activities and routines;

- > As labels on the bedroom door, toilet door, cupboards etc;
 - > To represent common places you may be going to when you leave the house, and attach these to the door you usually leave by; and
 - > As personal mementos in the bedroom that are placed on display, or in memory books and photo albums.
- Use strategies to help the person understand what you are saying:
 - > Get a person's attention before trying to communicate a message. Call them by their name, touch them gently on the arm, or both;
 - > Make eye contact;
 - > Always identify yourself;
 - > Use short complete sentences;
 - > Give one instruction at a time;
 - > Pause between instructions and bits of information;
 - > Use gestures like pointing to items or using a hand gesture to ask the person to stand;
 - > Show the person what to do; and
 - > Use touch based communication, such as touching the persons hand to prompt them to drink.

What happens to people with Alzheimer's disease over time?

Alzheimer's disease is a progressive condition over many years. Rapid changes in a person's abilities may indicate another problem, such as a chest infection or urine infection that requires prompt medical assessment and treatment.

In the more advanced stages of Alzheimer's disease the person may have physical problems such as:

- Incontinence;
- Difficulties with walking and falls;
- Difficulties with eating and swallowing, which may result in dehydration, malnutrition, choking and aspiration;
- Chest infections; and
- Seizures – may also occur in the early stages.

Alzheimer's disease progresses over a number of years. The rate of progression varies from person to person. However the disease does lead eventually to complete dependence and finally death, often as a result of chest infections or seizures. It is important to remember that with appropriate services and support, people with Alzheimer's disease can maintain quality of life.

It is most important to have a good working relationship with the person's general practitioner who can refer the person for specialist medical or allied health care if required, as well as make referrals to support services. For example if a person has difficulties swallowing then they should have a swallowing assessment by a qualified speech therapist as swallowing difficulties can result in repeated chest infections.

It is usually in the person's best interest for them to continue living in their home (including group home) for as long as possible. Home based supports are available for people with dementia and their carers.

Aged Care Assessment Services help people and their carers work out what kind of care will best meet their needs when they are no longer able to manage at home without assistance. Your general practitioner or local health service can refer you to an Aged Care Assessment Service, or you can contact the Commonwealth Carelink Centre. Alzheimer's Australia can also direct you to local support services.

There may come a time when the level of care required can no longer be practically or safely given in the home environment and the person may need to move into residential aged care. Alzheimer's Australia, Down Syndrome Australia or your own general practitioner can help you find information to assist you in making this decision.

Where can I get more information, advice and support?

Alzheimer's Australia

The Alzheimer's Australia website has lots of information regarding Alzheimer's disease, current treatments, support services, and information for family carers.

www.alzheimers.org.au

National Dementia Helpline

1800 100 500

Down Syndrome Victoria

Down Syndrome Victoria works to empower individuals to achieve a lifetime of meaningful inclusion in the community. Down Syndrome Victoria can also refer you to state organisations.

www.downsyndromevictoria.org.au

1300 658 873

The Centre for Developmental Disability Health Victoria (CDDHV)

www.cddh.monash.org

(03) 9501 2400

Commonwealth Carelink Centre

1800 052 222

Your local General Practitioner

Your local community health centre

Further reading

Understanding Learning Disability and Dementia; Developing Effective Interventions by Diana Kerr and Jessica Kingsley (2007)

Down's syndrome and Dementia by Diana Kerr (1997)

In the know; implementing good practice: information and tools for anyone supporting people with a learning disability and dementia by Diana Kerr and Heather Wilkinson (2005)

Contact details for State and Territory Down Syndrome Associations

Down Syndrome ACT

(02) 6290 0656

www.actdsa.asn.au

Down Syndrome NSW

(02) 9683 4333

www.dsansw.org.au

Down Syndrome NT

(08) 8985 6222

Down Syndrome QLD

(07) 3356 6655

www.dsaq.org.au

Down Syndrome SA

(08) 8369 1122

www.downssa.asn.au

Down Syndrome TAS

(03) 6224 0490

Down Syndrome WA

(08) 9358 3544

www.dsawa.asn.au



All photographs supplied by Mathew Wiggins. Mathew is a man living with Down syndrome, living well and realising his dream to become a professional photographer. For more information please email adm.red@bigpond.net.au.



Down Syndrome Victoria
www.downsyndromevictoria.org.au
1300 658 873

Alzheimer's Australia
www.alzheimers.org.au
National Dementia Helpline
1800 100 500

**The Centre for Developmental Disability
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