

Advancing Practice in the Care of People with Dementia

3rd edition

Module 9: Younger Onset Dementia



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Module 9: Younger onset dementia

Introduction

'Dementia' is the term used to describe symptoms of a large group of illnesses that cause a progressive decline in a person's mental functioning. It is a broad term that describes a loss of memory, intellect, rationality, social skills and normal emotional reactions.

The term 'younger onset dementia' is used to describe any form of dementia diagnosed in people under the age of 65. Although most dementias affect people who are elderly, occasionally younger people are diagnosed with dementia. Dementia has been diagnosed in people in their 50s, 40s and even in their 30s. Younger people tend to be affected by the less common forms of dementia, which often present with more challenging behaviours and progress at a faster rate (*Shunichiro et al., 2007*).

The previous Australian Institute of Health and Wellbeing's (AIHW) 2007 report on Dementia in Australia appears to have underestimated the number of people aged under 65 with dementia. The AIHW's report in 2012 produced statistics more in line with the way the World Health Organization (WHO) produces theirs and has estimated that 23,900 Australians aged under 65 had dementia in 2011, with men accounting for just over half; younger onset dementia represented about 8% of people with dementia in Australia. According to the WHO, younger onset dementia represents about 6–9% of all cases of dementia (AIHW, 2012).

Throughout this module we will present a case study to help demonstrate the unique challenges that the younger person with dementia and their family and carers face.

Objectives

On successful completion of this module you will be able to:

- Consider that younger people can have a diagnosis of dementia
- Describe the unique issues facing the younger person with dementia
- Understand the complexities of younger people accessing the aged care system
- Make links between Down syndrome and Alzheimer's disease

Module Topics

The complex process of diagnosis
 Types of younger onset dementia
 Reactions to diagnosis
 Relationships
 Practical issues

- Employment
- Financial

 Grief and Loss
 The role of counselling
 Access to relevant services
 Down syndrome and dementia
 References

Suggested reading for this module

Alzheimer’s Australia Website and Help sheets providing information on issues facing younger people with dementia:
<http://www.fightdementia.org.au/understanding-dementia/section-7-younger-onset-dementia.aspx>

Alzheimer's Australia. (2009). Quality Dementia Care Series No 5: Younger Onset Dementia – a practical guide. A report. Retrieved August 2014, from http://www.fightdementia.org.au/common/files/NAT/20090200_Nat_QDC_QDC5YODPracGuide.pdf

Alzheimer's Australia. (2008, revised 2013). Quality dementia care Series No 4: understanding younger onset dementia: A report. Retrieved August 2014 from http://www.fightdementia.org.au/common/files/NAT/20130808_QDC4.pdf

Alzheimer’s Australia: Websites and links, books, audio visual resources, journal articles, news stories available at:
<http://www.fightdementia.org.au/services/further-reading-and-resources.aspx>

Alzheimer's Australia. (2009). In our own words: younger onset dementia: a collection of very personal stories. Alzheimer's Australia: Canberra. Available at:
<http://www.fightdementia.org.au/common/files/NAT/20101027-Nat-YOD-InOurOwnWords.pdf>

Valenta, T. (2007). “Remember me. Mrs. V?” Caring for my wife: her Alzheimer’s and others’ stories. South Yarra: Michelle Anderson Publishing.
 Alzheimer’s Australia’s National helpline: 1800 100 500

The complex process of diagnosis

The process of diagnosing dementia can involve a number of health professionals, usually beginning with the general practitioner (GP), who will often refer the person to a specialist. Once referred to a specialist the person will undergo a series of tests, not only to diagnose dementia but also to determine the specific type of dementia. Cognitive screening and testing is usually performed to examine changes in a person's cognitive functions such as memory, orientation and planning. This is often done using the Mini-Mental State Examination (MMSE). Other tests such as blood investigations and structural and functional brain scans are also used.

For the younger person the road to a diagnosis of dementia can often be long and complex. It can take months or even years to obtain a correct diagnosis. Conditions such as depression, menopause, stress, other medical conditions and mental illness must first be ruled out. Extensive testing and delays in obtaining a correct diagnosis can lead to an increase in anxiety for the family while they await a correct diagnosis.

I was devastated; I couldn't believe it was Alzheimer's Disease. It had never crossed my mind, I just felt too young for something like that.

(Participant, AAV Younger Onset retreat. Marysville 2006)

An early diagnosis is essential to:

- Eliminate the possibility of other conditions that may have symptoms similar to dementia and may be treatable; these include depression, chest and urinary infections, vitamin deficiencies and brain tumours
- Eliminate other possible causes of confusion, such as poor eyesight or hearing, emotional changes, bereavement and loss, or side effects or interactions between certain drugs
- Allow early access to advice, information and support from Alzheimer's Australia, social services and support groups
- Allow the prescription of drugs that are effective in slowing the progression of certain types of dementia (e.g., Alzheimer's disease) in the early to middle stages, but are ineffective for other dementia types. An accurate diagnosis of the type of dementia is also important because drugs prescribed for other conditions could be detrimental to those with dementia (e.g., prescribing neuroleptics for mental health problems has very dangerous side effects for those with dementia with Lewy bodies)
- Allow the person with dementia and their family to plan and make arrangements for the future, such as writing a will, establishing an enduring power of attorney, writing advance care directives (a living will) and planning for financial management.

Alzheimer's Society UK (taken from In our own words)

Case Study

At the age of 44 Paul was a busy father, husband and a successful businessman. Paul had noticed that he was making mistakes at work. The mistakes were small at first but then became more frequent. Paul's wife Hanna also noticed changes. Paul seemed agitated and withdrawn from life. It wasn't until Paul's mother came to visit that the changes were really noticed and discussed.

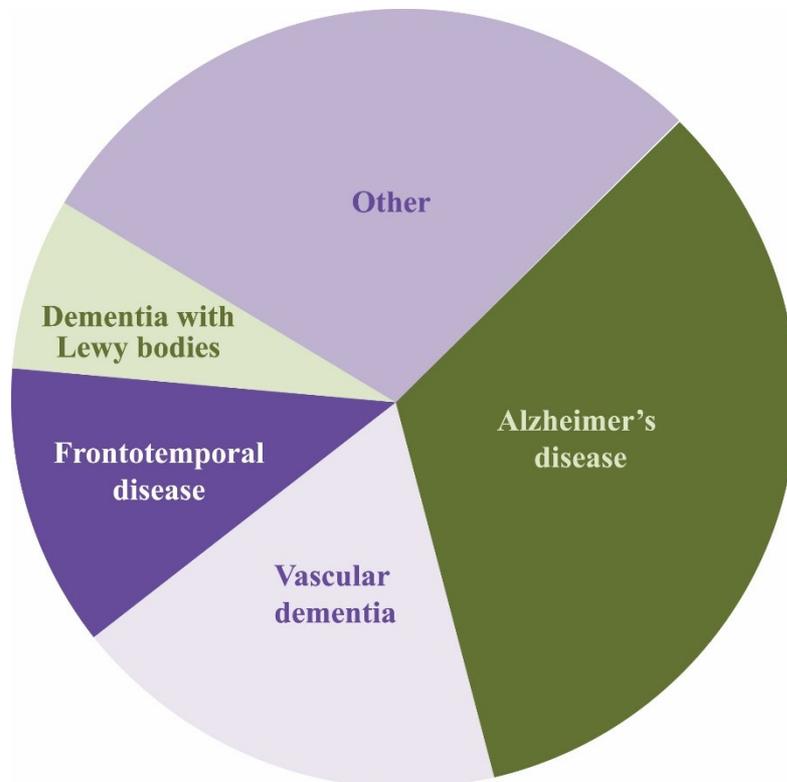
Paul and Hanna went to their local GP for advice. Paul was given a diagnosis of depression and was prescribed anti-depressants. Months passed but nothing had changed. Paul and Hanna went back to the GP, desperate for some answers to Paul's changes in mood and function. Hanna was advised to join a support group for people living with a partner with depression and Paul was encouraged to take some time off work. Another six months passed. Hanna was seeking some comfort from the support group but Paul was not improving.

On the third visit to the GP Paul was referred to a specialist for further testing.

The specialist diagnosed Fronto-temporal dementia. The diagnosis process took over 18 months and extensive testing during which time Paul, Hanna and their three children lived with the uncertainty of what was happening to Paul.

Types of younger onset dementia

Figure 1: Younger Onset Dementia comes in many forms



- Alzheimer's disease remains the most common form of dementia, even in adults under the age of 65 years.
- Younger onset Alzheimer's disease is more often inherited, especially when the onset is very early (40s or 50s).
- Up to half of the people with younger onset Alzheimer's disease have another family member with dementia.
- Much rarer is Familial Alzheimer's disease where the affected individual has two or more affected relatives (parent/grandparent).
- People with a diagnosis of Familial Alzheimer's disease should be offered genetic screening.

Hodges et al., 2009

Reactions to a diagnosis

A diagnosis of dementia under the age of 65 often comes as a shock. Dementia in the younger population is not well-understood and relatively rare within the community:

The biggest thing was accepting the diagnosis. It was especially difficult at such a young age. Linda (Alzheimer's Australia, 2009, p. 6)

Generally a diagnosis of younger onset dementia is not expected as dementia is often seen as a disease affecting older people.

The onset of dementia in someone of any age can bring many changes to the life of the individual, their family and friends. But when people are diagnosed with dementia at an early age, the challenges and issues are different. Younger people and their families have unique needs that must be considered. They may still be in the workforce or raising young families, or dealing with financial commitments and busy mid-life social relationships. In all cases the people are not old, but they now have a condition that is normally associated with older age, and they will be using a service system that does not fit their needs.

The diagnosis of dementia and the changes to memory and behaviour can have a huge emotional impact on the individual and the family around them. Feelings of shock, sadness, frustration, embarrassment, anger and loss are all common.

When the person diagnosed with dementia is younger, there can be an even greater emotional impact due to many varying reasons, including the shock of being so young, the greater impact on future life plans and the increased likelihood that children, financial responsibilities and employment may complicate the situation.

Every person's situation and the way they react are different, so it is expected that people will experience a wide range of feelings.

Case Study

Paul and Hanna had very different reactions to Paul's diagnosis of Frontotemporal dementia. Because the diagnosis process was so prolonged and complex Paul had undergone some significant deterioration in his condition. He is now at a stage where he cannot fully understand the impact of the diagnosis, nor does he have the capacity to contribute to future planning.

For Hanna the diagnosis was unexpected. Due to the rapid progression in Paul's condition Hanna had to take extended leave from work. Hanna also had the added complexity of trying to explain Paul's condition to their three young, school-aged children. For Hanna the changes have been many.

She now is responsible for everything to do with running the house, bringing up the children, trying to maintain the family business as well as struggling to make ends meet while dealing with the complexities of accessing Paul's superannuation and insurance payments.

Relationships

Family and friends

Younger onset dementia is a disease that affects the whole family and their social network. The carer and the person with younger onset dementia have to adjust to major shifts in their roles in the relationship. Carers may feel a sense of resentment that comes with the responsibility of looking after someone. Many people have to alter their hopes, dreams and expectations regarding their future together.

Carers often find the change of roles difficult to adjust to. For some families the person they are caring for is 'not the same person anymore'. Feelings of resentment, anger, guilt, frustration, love, hate, loss and grief are all normal (Hodges et al., 2009).

A younger person with a diagnosis of dementia is more likely to be actively raising a family, thus leaving the household severely disrupted. The impact on the family may be far greater than that of an older person, with large emotional, financial and social impacts on the immediate and extended family.

Families and carers can often have a difficult time caring for someone when they are also engaged in a busy life; they may still have children at home or elderly parents requiring assistance. For many partners the diagnosis will have a dramatic impact on their lifestyle:

Your life seems to change overnight. My partner had to leave work pretty much after the diagnosis as I needed his support at home. We are relatively happy with life now and we just had to leave our precious life behind us.

(Participant, Alzheimer's Australia, Vic. Younger Onset retreat. Marysville 2006)

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(Participant, Alzheimer's Australia, Vic. Younger Onset retreat. Marysville 2006)

For most children the diagnosis of a progressive chronic illness in one of their parents is devastating. Children are very vulnerable and are likely to have difficulty understanding the illness in their parent and the inevitable change in roles within the family. They may not understand the changes in behaviour that happens in dementia and may think that they are causing the behavioural problems. They may benefit from counselling to assist with many conflicting emotions:

As a sixteen-year-old boy being told this about your father, who in your eyes is Superman, completely bullet-proof and immortal, was something I wasn't ready to deal with. This was the man that could teach you anything and fix anything for you and now they're helpless to help him.

(Valenta, 2007)

Case Study

For Paul and Hanna relationships changed greatly. Hanna's role of a wife and mother quickly became a role that needed to encompass every element, from running the household, taking over the family business and raising the kids. Hanna also had to give up her university studies and paid employment to focus on the needs of her family.

Hanna's mother took on a greater role in caring for her son-in-law as well as supporting her grandchildren.

Paul and Hanna's intimate relationship also changed as the illness progressed. They could no longer share the same bed or have the same meaningful conversations that once represented their relationship.

The older children took on a larger role in caring for their father and supporting their mother in the day-to-day running of the family business. This had an impact on their ability to do the things that kids typically do, such as participate in sporting clubs. The family also began to struggle financially, making extra-curricular activities for the children unaffordable.

Practical Issues

Employment

Often the younger person is still in the workforce when symptoms of dementia are noticed or when the diagnosis is made. This can have a huge impact on the family for many reasons:

- Work performance may become affected and progressively have more impact.
- The decision to tell or not to tell an employer about the diagnosis needs to be discussed.
- Professional advice may be required to ensure that the best financial decisions are made in relation to superannuation, insurance entitlements and other financial considerations.
- Discussing plans around the continuation of employment and the impact of early retirement are difficult. People may feel a sense of loss when their plans for the future are altered.

Early, unexpected retirement can have a huge impact on the individual and family. A younger person with dementia should seek advice if they have left or are planning to leave employment to ensure that they are accessing all their entitlements to superannuation and insurance benefits. The person and their family may require assistance and support from health professionals to make these adjustments.

Financial

People with dementia can often have difficulties performing activities that were once very easy. Breakdown in the ability to manage personal and family finances can be due to a lack of motivation and interest, memory loss, language difficulties, or planning and organisation problems.

Other financial implications are many and varied. Financial challenges can become evident with the younger person with dementia not being able to continue to work, or for their partner in having to reduce or leave work in order to care for the person with dementia. Full-time carers may be eligible for the Carer's Payment or Carers Allowance.

Case Study

Paul has his own business which had begun to fail as a result of his then undiagnosed condition. The family now have the challenge of building up and maintaining the business as it is their main source of income. Thankfully Hanna and Paul had some years earlier put Powers of Attorney in place, which has enabled Hanna to access business accounts immediately.

The financial strain of Paul's diagnosis has been profound. The family are unable to afford the mortgage repayments on the house and are looking at their options for downsizing. Hanna has found that attempting to access Paul's insurance benefits has been a very long and complex process at a time when they need access to the funds.

Grief and Loss

There is often a strong sense of grief and loss experienced by younger people with dementia and their families. For younger people, grief seems to be more profound than for older people, as their sense of loss can be greater. This comes from having to engage in the changes in their lives and the likelihood they will not be able to carry out their plans for the future. Some of the loss that may be experienced can include:

- Relationships with a spouse or partner will change; the partnership enjoyed intimacy and shared dreams and there is often greater strain placed on relationships
- Loss of independence and increasing dependence
- Loss or restriction of driving licence
- Loss of life expectations.

It is so hard to watch my partner, someone I love go through this. I was at the movies the other day (thankfully by myself) and I burst into tears and could not stop crying. When I stopped crying I realised I was crying for all of us.

(Carer Participant. AAV Younger Onset retreat. Marysville 2006)

Driving

The decision to give up driving because of dementia is difficult at any age. Driving allows us the independence and autonomy to have control in what we do and where we go.

Many young people with dementia speak of the difficulty in giving up driving when the onset of dementia was unexpected. Those with children can no longer drive their children to school, further isolating them from playing an active role in the children's upbringing. Some younger people have expressed feelings of guilt at the thought of letting their children down (Alzheimer's Australia, 2007).

There are limited alternative transport options available for younger people with dementia. There can be issues in accessing the Multi-Purpose Taxi Program, which allows card-holders half-price taxi fares. The program is means-tested, which often results in younger people being denied access if their partner is still actively employed. This often results in the person with dementia becoming more isolated, especially while their partner is working.

By law, it is the person with dementia or the family's responsibility to report to the Driving Licensing Authority any permanent or long-term illness that is likely to affect the ability to drive safely. A driving assessment can be conducted to determine if a person is able to continue driving, even with a restricted licence.

The role of counselling

Counselling offered by health agencies and organisations such as Alzheimer's Australia aim to provide information, advocacy and support. Time is allowed for the person and their carer to explore their experience of dementia as well as the more pragmatic aspects of managing the practicalities of the situation. Alzheimer's Australia (Vic.) offers many services to the younger person with dementia and their family and carers. Counselling, support, advocacy and education are all available. People in the early stages of the illness also have access to the Younger Onset Dementia Living with Memory Loss program, providing an opportunity to access information, advice and peer support:

It is a very useful experience and it helps greatly to be able to talk to people who understand our situation and problems

(Carer Participant. AAV Younger Onset retreat. Marysville 2006)

Case Study

Paul and Hanna have both accessed support from Alzheimer's Australia, Vic. and other organisations. The focus for Hanna has been around accessing supports for the children and counselling for herself. Both Paul and Hanna have attended a Living with Memory Loss group.

Access to relevant services

Although approximately 23,900 people are living with younger onset dementia in Australia, appropriate services are limited. For the most part, people with younger onset dementia are accessing a service system that does not fit their unique needs. Typically, the younger person with a diagnosis of dementia accesses the aged care system of support; this doesn't address their younger needs, such as having children at home and being physically fit and active.

As the dementia progresses and the person needs a higher level of care, permanent aged care may be considered. Currently few facilities exist in Australia that can meet the needs of the younger person in a nursing home, which is predominately made up of the older population.

Case Study

Hanna has commented that there were many supports available to them when they had a diagnosis of depression; however, once the diagnosis changed to dementia the services available to them that were age-appropriate were limited. Paul currently attends a Planned Activity Group once a week; however, he is the youngest group member by 17 years. He is the only member of the group that is physically fit and therefore is restricted in the types of activities that he can access. Due to Paul's rapid deterioration, Hanna is looking at residential care options for Paul and is struggling with the thought that he will need to enter an Aged Care facility. This makes the already difficult process of considering residential harder for the younger person

Down syndrome and Alzheimer's disease

The link between Down syndrome and Alzheimer's disease is well-documented (Kimura et al., 2007; Nelson, Scheibel, Ringman, & Sayre, 2007). Studies show that by the age of 40, almost 100% of people with Down syndrome who die have the changes in the brain associated with Alzheimer's disease. Amyloid precursor protein (APP—the abnormal breakdown that yields the toxic amyloid protein which forms plaques in the brain and probably damages brain cells and their connections—is coded for chromosome 21.

People with Down syndrome have an extra copy of chromosome 21 and produce 1.5 times as much APP as other people; this seems to result in an excess tendency for the abnormal amyloid breakdown product to build up, and seems to cause earlier appearance of the brain changes typical of Alzheimer's disease.

However, a significant number of people with Down syndrome are older than 40 and show no signs of having Alzheimer's disease. It is not currently understood why changes to the brain that are typical of Alzheimer's disease do not necessarily produce the condition in people with Down syndrome.

Summary

Dementia is generally seen as a disease of older age. This module has identified that younger people may also be living with dementia. The case study provides a working example of the complexities of the illness in the younger population and explains some of the difficulties the younger person has in accessing relevant and appropriate services.

Video

Website and Information

Resources

Robertson, A., & Rogers, P. (2000). Video: *What is happening to my friend Mary?* The Lorna Hodgkinson Sunshine Home.

Exploring the issues of Down syndrome and Alzheimer's disease: Centre for Developmental Disability Health Victoria. Fact sheet Retrieved August 2014, from <http://www.cddh.monash.org/assets/fs-downsynd.pdf> and Booklet *Down Syndrome and Alzheimer's Disease* <http://www.cddh.monash.org/assets/dsad-booklet-final.pdf>

Centre for research on families and relationships.

Research briefing 17: *Growing older with a learning disability and dementia* Retrieved August 2014 from <http://www.era.lib.ed.ac.uk/handle/1842/2820>

Research briefing 28: *Best practice in learning disability and dementia* Retrieved August 2014 from <http://www.era.lib.ed.ac.uk/handle/1842/2781e>

Alzheimer's Australia Website and Help sheets provide information on issues *Down syndrome and Alzheimer's disease* Retrieved August 2014 from

<http://www.fightdementia.org.au/understanding-dementia/down-syndrome--alzheimers-disease.aspx>

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Access Economics. (2011). *Dementia Across Australia 2011–2050*. Retrieved August 2014 from

http://www.fightdementia.org.au/common/files/NAT/20111014_Nat_Access_DemAcrossAust.pdf

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Nelson, L.D., Scheibel, K. E., Ringman, J. M., & Sayre, J. W. (2007) An experimental approach to detecting dementia in Down syndrome: A paradigm for Alzheimer's disease. *Brain and Cognition*, *64*(1), 1-110.

Valenta, T. (2007). In the name of my father. In *"Remember me. Mrs. V?" Caring for my wife: her Alzheimer's and others' stories*. South Yarra: Michelle Anderson Publishing Pty. Ltd.

Kimura, R., Kamino, K., Yamamoto, M., Nuripa, A., Kida, T., Kazui, H. et. al. (2007). The DYRK1A gene, encoded in chromosome 21 **Down syndrome** critical region, bridges between beta-amyloid production and tau phosphorylation in Alzheimer disease. *Human Molecular Genetics*, *16*(1), 15-23.

Shunichiro, S., Manabu, I., Yasutaka, T., Teruhisa, M. et al. (2007). Frequency and clinical characteristics of Early-Onset Dementia in consecutive patients in a memory clinic. *Dementia and Geriatric Cognitive Disorders*, *24* (1), 42.