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In this article...

- The types, prevalence and diagnosis of young-onset dementia
- The needs of the person affected, as well as their family members and dependents
- Why young-onset dementia is often unrecognised, misdiagnosed or underdiagnosed

Dementia 2: diagnosing young-onset dementia and supporting patients

Key points

Nurses in all settings should be aware of the signs and symptoms of young-onset dementia

Long delays in diagnosis can result in people and their families trying to cope without support

Access to high-quality diagnostics leads to improvements in wellbeing for people living with young-onset dementia and their families

Routine support, such as day care, is often focused on the needs of older people with dementia

Authors Hannah Gardner is Admiral Nurse, Admiral Nurse Dementia Helpline; Amy Pepper is Admiral Nurse research assistant, Dementia UK.

Abstract In the UK, young-onset dementia accounts for a smaller number of people living with dementia than those with later-onset dementia, but the number is still significant. People diagnosed with young-onset dementia face a number of unique challenges, including managing a life-limiting condition while in employment or having caring responsibilities for children, managing finances and accessing age-appropriate services. It is important that nurses are aware of young-onset dementia and how best to support families living with it. This second article in a series on dementia considers the causes of young-onset dementia, the assessment process, the implications for families living with the condition, and outlines recommended care and support.

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The high prevalence of dementia in older people can overshadow the importance of its occurrence in younger people (Rossor et al, 2010). An estimated 7.5% (70,800 people) of the estimated 944,000 people living with dementia in the UK are living with young-onset dementia (YOD), where symptoms occurred under the age of 65 (Carter et al, 2022). The estimated prevalence figure for YOD in England, where diagnosis was between the ages of 30 and 64, is 92 per 100,000 of the general population (Carter et al, 2022).

The cut-off point of 65 years has largely been sociologically driven by:

- This being the standard age, in the UK, for the transition from employment to retirement;
- The fact that health and social care services are delivered by age, such as in old-age psychiatry and geriatric medicine.

However, the age of 65 years has no specific biological significance and some disease features are shared across both age groups, despite this arbitrary divide (Rossor et al, 2010).

Types of YOD

YOD can be caused by a variety of aetiologies (Box 1). Alzheimer's disease is the most common cause of YOD, but the proportion of cases of YOD caused by Alzheimer's disease is smaller than that seen in later-onset dementia (Draper and Withall, 2016).

Other prominent causes of YOD include:

- Frontotemporal dementia;
- Huntington's disease.

Both of these occur more frequently in YOD cases than in later-onset dementia cases (Rossor et al, 2010).

With growing alcohol use among younger and middle-aged populations globally, alcohol-related dementia is likely to become more common (Davis et al, 2022). Regularly consuming alcohol in large quantities can cause alcohol-related brain damage and result in:

- Wernicke's encephalopathy;
- Korsakoff syndrome;

- Diffuse alcohol-related brain damage;
- Alcohol-related amnesia (Davis et al, 2022).

As a result of the large variation in the causes of YOD, there are a range of presenting symptoms. A relatively large proportion of YOD cases are caused by Alzheimer's disease but, often, their presentation in younger people can be atypical, including problems with language, visual-spatial skills and executive function (Graff-Radford et al, 2021); however, the more-typical cognitive symptoms can also occur, such as memory problems (Draper and Withall, 2016).

Depression is also a common early feature of YOD (Rossor et al, 2010) and, due to the over-representation of frontotemporal dementia in people with YOD, there can also be early behavioural and personality changes, including problems with:

- Empathy;
- Disinhibition;
- Social withdrawal (Draper and Withall, 2016).

YOD is still rare, but is also more likely to be inherited than later-onset dementia – for example, inherited cases account for 10% of all people diagnosed with young-onset Alzheimer's disease (Mol et al, 2022). In families that have an inherited condition, genetic counselling and testing may be available (Loy et al, 2014).

Assessment and diagnosis

An overview of the assessment process, which includes tests of mental abilities and at least one brain scan, is outlined in Box 2. Despite the advancement of methods of diagnosis, YOD is often underdiagnosed or misdiagnosed (Kuruppu and Matthews, 2013); in addition, in some cases, people may receive an incorrect diagnosis of YOD due to lack of understanding and resources to educate health professionals (O'Malley et al, 2021). People living with YOD wait an average of just over four years for a diagnosis – almost double the time for people aged >65 years (van Vliet et al, 2013).

Delays to diagnosis can be attributed to a number of factors:

- YOD is relatively rare, so clinicians may not attribute symptoms to dementia;
- The early symptoms of YOD may differ from those of dementia occurring after the age of 65 years;
- Diagnosis is often managed in settings geared towards older people living with dementia, in which the specialist knowledge needed to effectively diagnose YOD may be lacking (Ray and Dening, 2021).

These long delays in diagnosis can leave people living with YOD and their families trying to cope without support, during which time their condition may worsen, which can place strain on the whole family unit (Young Dementia Network, 2022).

Box 1. **Young-onset dementias**

Frontotemporal dementia
Umbrella term for a group of dementias that mainly affect the frontal and temporal lobes of the brain. Frontotemporal dementia tends to occur in people aged 45-65 years, and accounts for around 12% of all young-onset dementia diagnoses

Lewy body dementia
Appears to be relatively uncommon in younger populations, with a clinical presentation similar to that seen in older people

Alcohol-related brain damage and dementia
Caused by drinking alcohol excessively over a prolonged period and by a combination of factors, including vitamin B1 (thiamine) deficiency, the toxic effects of alcohol on nerve cells, head injury and blood-vessel damage. Alcohol consumption is rising rapidly globally in young and middle-aged people, who are placed at risk of developing alcohol-related brain damage and dementia

Posterior cortical atrophy
Caused by degeneration of the cells at the back (posterior) of the brain. Has similar pathology to Alzheimer's disease, but early symptoms will involve sight, and problems with spelling, writing or maths

Huntington's disease
Genetic disorder caused by a faulty gene on chromosome 4; results in problems with movement, learning, cognition and emotions

Alzheimer's disease
The most common cause of young-onset dementia. A small number of people have an inherited form called familial Alzheimer's disease, which typically develops before the age of 65 years and, most commonly, in people who are in their 40s or 50s

Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL)
A rare genetic form of vascular dementia symptoms, including migraine, repeated stroke, loss of mental abilities, seizure, apathy and depression. Onset is usually in the person's mid-30s and inherited from a parent, who has a 50% chance of passing on the mutated copy of the gene

Vascular dementia
Caused by problems in the blood supply to the brain and closely linked to both diabetes and cardiovascular diseases, such as stroke and heart disease. Symptoms vary from person to person. Early memory loss is less common than in other young-onset dementias, but other symptoms are more common; these include slower speed of thought and problems with planning, organising, making decisions, solving problems, following a series of steps and concentrating

Source: Adapted from Gerritsen (2020)

"People diagnosed with young-onset dementia face unique challenges, many of which differ from those faced by people living with dementia at an older age"

In some areas of the UK, specialist YOD services provide assessment and post-diagnostic support but these are not available to everyone (Stamou et al, 2022; Rayment and Kuruvilla, 2015). A specialist approach is needed, particularly given the higher incidence of rarer types of dementia in people with YOD (O'Malley et al, 2019). The National Institute for Health and Care Excellence (NICE) (2018) recommends specialist investigations including:

- In-depth imaging;
- Neuropsychological testing;

People with young-onset dementia and their family members can find it difficult to access specialist support services

- The use of biomarkers and genetic testing (measures of what is happening inside the living body, shown by the results of laboratory and imaging tests).

We know that access to high-quality diagnostics leads to improvements in well-being for people living with YOD and their families/carers (Stamou et al, 2021). Despite this, a recent analysis showed that an acceptable diagnostic procedure was followed in just 24% of cases (Stamou et al, 2021), and people living with YOD themselves have highlighted this as an area that needs improvement (O'Malley et al, 2021).

Case management

People living with YOD benefit from a multidisciplinary team approach to their diagnosis and care, with shared working across specialities, along with key workers or case managers who can refer to age-appropriate services with appropriate activities (Stamou et al, 2021).

Dementia UK has developed a range of resources to support families affected by YOD. In addition, in 2020, it merged with the charity YoungDementia UK to bring together information and resources created specifically for YOD and cover the key issues that families and carers face. This year, together, the charities have launched a consultant Admiral Nurse role, a nurse with specialist knowledge and skills to meet the needs of families affected by YOD, given their often unique set of circumstances that differ from those with later-onset dementia.

Admiral Nurses are specialists in dementia care who, through a case-management approach, work across health and social care systems to deliver clinical support to families affected by dementia who have complex needs. The Admiral Nurse model aims to deliver case management across the life course of dementia (Harrison Dening et al, 2017), from peridiagnosis to end of life, as well as bereavement support for family members.

Box 2. Assessment process

- Full history of the person's symptoms, any changes they have experienced and the impact of these on day-to-day activities
- Tests of a person's mental abilities, behaviour and ability to do daily tasks
- A full physical examination
- At least one brain scan (often with more specialist scans such as a positron emission tomography scan)
- Review of other health conditions and how these can be managed effectively
- Lumbar puncture to collect and analyse fluid around the spine

Living with YOD

People diagnosed with YOD face unique challenges, many of which differ from those faced by people living with dementia at an older age. They may have caring responsibilities and their being of working age can mean that employment and finances are affected, as well as social and spousal relationships (Young Dementia Network, 2022).

People with YOD may find it harder to access age-appropriate support for themselves and their families/carers, leaving families affected feeling isolated. A recent report from the Young Dementia Network (2022) looked at how NICE's guidance for dementia could be adapted to better suit the needs of people living with YOD. It highlighted five areas including:

- Involving people living with YOD in decisions about their care;
- Diagnosis;
- Care coordination;
- Interventions to promote wellbeing and cognition;
- Support for carers.

There are several specific interventions that should be considered for families living with YOD, covering issues such as:

- Financial planning and support;
- Psychosocial activities;
- Behavioural symptoms and their management;
- Support for family carers, including children.

Financial planning and support

People living with YOD may still be in employment or have a mortgage or other

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debts, or financial dependants in the form of young children. As such, their needs may be quite different from people aged >65 years who are living with a dementia. Recent research by Mayrhofer et al (2021) into the financial consequences of a diagnosis of YOD found that, for many people:

- Financial hardship occurred as a direct consequence of the diagnosis;
- Not only can there be a loss of income due to early retirement, but also a loss of subsequent pension contributions.

There are a number of potential avenues for financial support after a diagnosis of YOD, including:

- Personal Independence Payment;
- A social care needs assessment and associated financial assessment, used to assess how much someone will have to contribute towards their care;
- Carers' assessment;
- NHS continuing healthcare.

However, these systems and how to apply for them are often poorly understood by families living with YOD (Mayrhofer et al, 2021).

Many of those surveyed had to go through the appeals processes to obtain benefits to which they were entitled; in some cases, they were only successful when specialist support was given to support the appeals process (Mayrhofer et al, 2021). There is a clear need for families living with YOD to be given, not just information and advice about managing their finances and applying for benefits, but also detailed support to help them navigate the practicalities and emotional impact.

Psychosocial activities

People living with YOD should be offered opportunities to access meaningful and age-appropriate activities. Non-pharmacological interventions, tailored to the person, should be offered and can include:

- Cognitive stimulation therapy;
- Physiotherapy;
- Occupational therapy (NICE, 2018).

People living with YOD – particularly those primarily affected by language difficulties – could benefit from speech and language therapy (SLT). Several studies have shown that word-retrieval interventions can be beneficial in people with YOD, and especially with primary progressive aphasia (PPA) (difficulty expressing thoughts and understanding or finding words) (Jokel et al, 2014).

However, there is a limited availability of SLT services in YOD (Rayment and Kuruvilla, 2015) and, when a referral is made to a generic SLT service, many SLT practitioners lack the knowledge and skills to support people with YOD, and especially PPA. As with many services specific to YOD, specialist PPA SLT services are rare and unevenly distributed across the UK (Volkmer et al, 2020).

Peer support groups can be helpful, but many dementia support groups may be unsuitable for people living with YOD. A lack of common interests with older people and a wish to socialise with people of a similar age have been mentioned as factors influencing service attendance (Rabanal et al, 2018). People with YOD are often reluctant to attend day centres that largely meet the needs of people with dementia who are aged >65 years (Carter et al, 2022).

> *"For many people, financial hardship occurred as a direct consequence of the diagnosis"*

Depression and anxiety are common in people living with dementia, but a recent study by Bell et al (2022) found that, although talking therapies (psychological treatments for mental and emotional problems, such as stress, anxiety and depression) were of some benefit in people with dementia and YOD, they were not as effective as they were for people without dementia. The authors call for more research to improve access to psychological therapies and facilitate a better understanding of how such therapies can be adapted to further improve outcomes for people with dementia.

Behavioural symptoms and their management

Changes in behaviour are common in YOD, across the various subtypes (Sansoni et al, 2016); these can lead to relationship breakdown and increase the risks of premature admission to residential care (Rasmussen et al, 2019). These changes can take many forms, but can be broadly categorised as:

- Aggression;
- Compulsive behaviours;
- Disinhibition;
- Socially inappropriate behaviours (O'Connor et al, 2022).

A recent study by O'Connor et al (2022) found a high incidence of behavioural changes reported by families living with YOD and recommended specialist support services for this group of people; these should include education and support for family carers in how to manage these types of symptoms. Despite the clear need for such support, the availability of services tailored to those living with YOD in the UK has been reported as being patchy (Rayment and Kuruvilla, 2015). Where there are no specialist YOD services available, support may be offered via local memory services, Admiral Nurse teams or through voluntary services such as Dementia UK and the Young Dementia Network.

In some cases, medications may help in the treatment of behavioural symptoms, but these should always be used as a last resort and following specialist assessment. The next article in this series will explore in more detail medical treatments for dementia, including medications.

Support for family carers, including children

People with YOD may have dependent children and/or be carers themselves for their own older parents. There is a stereotypical view that dementia is an older person's disease, which means the specific needs of a young child of a person with YOD can be overlooked (Blake and Hopper, 2022). This can lead to children and young adults being affected by a lack of awareness and stigma, resulting in significant psychosocial problems (Blake and Hopper, 2022).

The children of a person living with YOD can often assume caring responsibilities for their parent and will have very different needs to other carers of people with dementia, especially those who are likely to be adults caring for an older parent or grandparent with later-onset dementia (Sikes and Hall, 2018).

Children should be given young carer support through school, and teachers need to seek resources and training to understand the impact of YOD on children (Felc et al, 2021). There are several resources and websites that aim to support young carers to cope with their caring role, including Carers Trust (nd) and NHS (2021). However, many children will require more direct guidance and support, so schools and health and social care organisations should ensure the needs of these children are appropriately recognised and assessed to enable access to practical, psychological and emotional support (Young Dementia Network, 2022).

Conclusion

Living with YOD brings challenges and hurdles that are very different to those experienced in later-onset dementia. Often families affected feel isolated and early symptoms are dismissed or diagnosed as other conditions, such as depression, menopause or even anxiety. With the increase in

"The children of a person living with young-onset dementia can often assume caring responsibilities for their parent"

people developing dementia, nurses need to be more aware of YOD, its impact on the whole family and the challenges they face, especially in getting a diagnosis and access to age-appropriate support.

Equally challenging, once an accurate diagnosis has been made, are conversations about future care. Future decision-making for families affected by YOD can be difficult due to the individual receiving a diagnosis of dementia in midlife, potentially having young dependents and the financial pressures imposed by still being of working age. By increasing the awareness of YOD, we can support younger people who are worried about symptoms that are indicative of dementia to visit their GP in a timely manner, thereby facilitating early access to appropriate support and treatment. **NT**

● The third article in this series will consider the medical treatment and management of dementia.

Useful online resources

Dementia UK
Young onset dementia
dementiauk.org/about-dementia/young-onset-dementia

Young Dementia Network
A community of people living with young onset dementia, their family and friends, as well as professionals in health and social care and the voluntary sector
youngdementianetwork.org

Rare Dementia Support
Information and support for people affected by, or at risk of, a rare dementia
raredementiasupport.org

Social Care Institute for Excellence
Young onset dementia: services and support
scie.org.uk/dementia/symptoms/young-onset/support-for-younger-people.asp

Alzheimer's Society
Services for people with young-onset dementia
alzheimers.org.uk/about-dementia/types-dementia/services-people-young-onset-dementia

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