Factsheet 1

Young Onset Dementia (YOD)

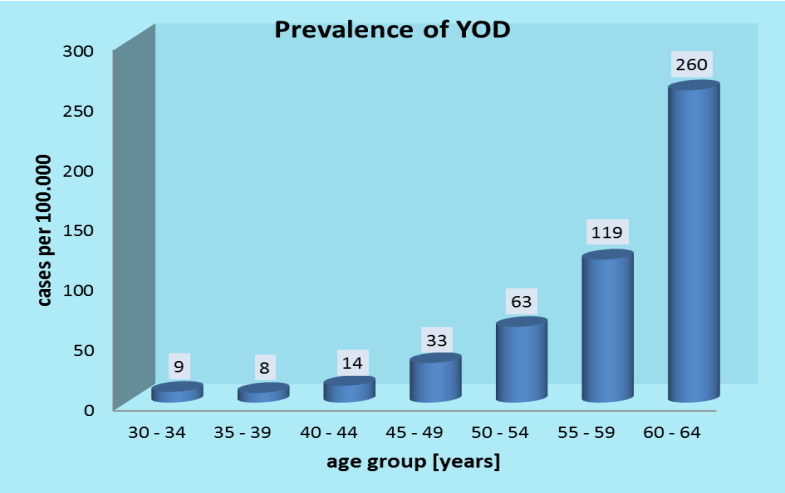
Dementia is commonly seen as a health and social problem of older adults. Nevertheless dementia can occur earlier in life. Young onset dementia is defined by an onset of symptoms before the age of 65 years.

When dementia starts at a relatively young age it is associated with specific and distinctive needs for those with the diagnosis, relatives and healthcare professionals.

How many people are affected?

The frequency of YOD is estimated at 100 per 100,000 in the population aged between 45 and 65 years while the prevalence of dementia in older population is about 5,000 in 100,000. The rate of new cases of YOD is 5 - 20 per 100,000 per year in this age group.

The prevalence of YOD increases almost exponentially with age (as does the prevalence of late onset dementia).

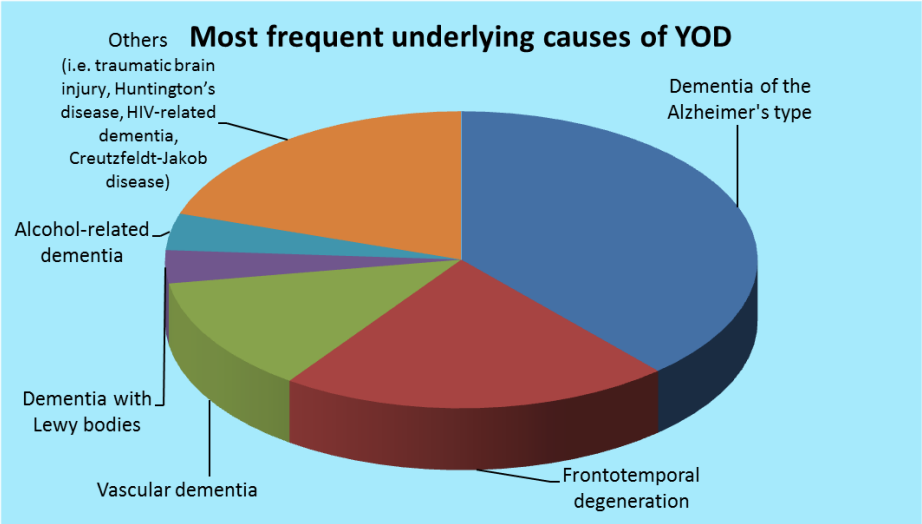


Source: Vieira et al.: Clin Pract Epidemiol Ment Health 9: 88–95, 2013

Causes of dementia in younger people

The cluster of symptoms that we call dementia can be caused by a number of different diseases of the brain. The most frequent underlying causes of YOD are diseases in which nerve cells are damaged and gradually become dysfunctional (Alzheimer's disease, Frontotemporal degeneration) or where nerve cells are lost due to lack of blood supply as a consequence of small vessel occlusions (Cerebrovascular diseases). Genetic causes play a far greater role in YOD than in late-onset dementia.

Alzheimer's disease is the most common form of dementia in younger people and a third of younger people with dementia have Alzheimer's disease. However, this is a far smaller proportion than in older people with dementia, where around two-thirds of people have Alzheimer's disease.



Source: Shinagawa et al.: Dement Geriatr Cogn Disord 2007

Alzheimer's disease

Alzheimer's disease causes two characteristic changes in the brain tissue. Firstly, senile plaques are deposited outside of nerve cells. These are composed of a protein called amyloid that has been abnormally processed. Secondly, neurofibrillary tangles are formed from a protein called tau that also undergoes a change in composition causing it to clot together within nerve cells. Senile plaques and neurofibrillary tangles are found in abundance in affected brain regions. Both pathological changes cause nerve cells to become dysfunctional and ultimately disappear.

At the early stages of the disease memory problems mainly refer to the ability to learn new information. Memory of past events is usually preserved until a later stage of the disease. Additional symptoms include reduced attention and word-finding difficulties. Executive functions, such as understanding situations, taking decisions, planning and organising, are also compromised. The ability to orientate oneself in time and space usually gets affected at the moderate stage of the illness.

Alzheimer's disease is also associated with a range of behavioural changes. At the early stage, depression and apathy are most frequent. At the moderate stage, agitation and irritability may occur. Later on, people may get day mixed up with night, become aggressive, shout or fidget.

In approximately one third of patients with young onset Alzheimer's, the disease has an atypical appearance. In a frontal variant, behavioural changes occur that are very similar to those seen in the behavioural variant of Frontotemporal dementia. There is also a language-dominant form with severe word-finding difficulty and slowing of speech. The third variant is characterised by particular problems with vision.

Frontotemporal degeneration

The Frontotemporal degenerations are a group of brain diseases which primarily affect the frontal lobes and the anterior parts of the temporal lobes. They are varying in terms of pathology, genetics and clinical appearance. Together, they represent the second most frequent cause of YOD. Frontotemporal degeneration most commonly occurs between the ages of 50 and 60. In about 40 percent of cases, the person has a family history of dementia.

The Frontotemporal degenerations have three distinct clinical appearances that depend on the localisation of the disease process:

Behavioural variant Frontotemporal Dementia is characterised by early behavioural changes including disinhibition, loss of tactfulness, apathy, inertia, impulsivity, loss of empathy, repetitive or stereotyped behaviours, and altered dietary habits. These are often interpreted as an alteration of personality. There is also cognitive impairment, although this is less prominent than in Alzheimer's disease. It mainly involves executive functions, such as planning and organising, while memory is relatively spared. Due to the combination of behavioural changes and executive dysfunction more complex activities of daily living are rapidly lost. It is very unlikely that the affected person is aware of problems.

In **non-fluent variant Primary Progressive Aphasia** people have increasing trouble producing speech. They have a slow and halting speech. However, the ability to understand language is preserved. Over time, individuals invariably show behavioural changes similar to those in Behavioural variant Frontotemporal dementia.

Semantic-variant Primary Progressive Aphasia does not primarily affect language production but language comprehension. People are no longer aware of the meaning of words. Later they also have difficulty recognising names and familiar faces. Although they are able to speak fluently, their communication tends to have poor content and meaning. In the course of the illness individuals also develop behavioural changes as for those seen in Behavioural variant Frontotemporal dementia.

Some people with Frontotemporal degeneration develop motor symptoms of Parkinson's disease, including rigidity, slow movement and balance problems. Others experience signs of motor neuron disease which is also called Amyotrophic lateral sclerosis.

Vascular dementia

Vascular dementia is caused by disease of small blood vessels within the brain. Narrowing and occlusion of these vessels reduces the blood supply, particularly in the deep parts of the brain. This results in a localised loss of brain tissue including nerve cells. If a large area is affected this is called an infarct, if the area is small it is referred to as lacuna. Another consequence of reduced blood supply is a widespread damage to nerve fibres which connect the different brain regions.

This death of brain cells can cause problems with memory, thinking or reasoning. The way vascular dementia affects people varies. Symptoms may develop suddenly or more gradually.

Memory loss is not usually the main symptom in the early stage of vascular dementia. The most common cognitive symptoms in the early stages of vascular dementia are: problems with planning or organising, making decisions or solving problems; difficulties following a series of steps (for example cooking a meal); slowing of thought; and problems concentrating, including short periods of sudden confusion. A person in the early stages of vascular dementia may also have difficulties with memory: problems recalling recent events (often mild); language (for example, speech may become less fluent); and visuospatial skills (such as problems perceiving objects in three dimensions).

When dementia is caused by a combination of Alzheimer's disease and cerebrovascular damage, this is called "mixed dementia". In older individuals this may be the most common cause of dementia. In younger individuals, mixed dementia is rarer.

Dementia with Lewy bodies

Dementia with Lewy bodies includes Parkinson's disease dementia. Pathologically it is characterized by the development of abnormal collections of protein within the cytoplasm of neurons (known as Lewy bodies). Symptoms of dementia with Lewy bodies include problems with confusion and alertness fluctuating significantly throughout the day. People usually develop typical Parkinsonian movement abnormalities (such as slower movements, postural instability or tremor) and vivid visual hallucinations. Also typical is a problem known as rapid eye movement (REM) sleep disorder where people act out dreams – sometimes with violent movements. Memory loss may be significant but less prominent than in Alzheimer's disease.

Other neurological disorders

Unlike the condition in older individuals, dementia in younger people is more often caused by rare neurological disorders. These include traumatic brain injury, Huntington's disease and normal pressure hydrocephalus.

Infectious diseases

Infectious diseases of the brain are rare causes of dementia in younger people. They include Creutzfeldt-Jakob disease and HIV-related dementia.

Alcohol-related dementia

Alcohol-related dementia can occur in people who have regularly consumed large volume alcohol over extended periods of time. It should be distinguished from Wernicke-Korsakoff syndrome, which is also caused by excessive and prolonged alcohol use but alcohol-related dementia predominantly affects memory. This form of dementia could be due to both the toxic effects of alcohol on nerve cells and the destructing and lasting impact of thiamine (vitamin B1) deficiency on the nervous system. Less than 10 percent of dementias in younger people are alcohol related.

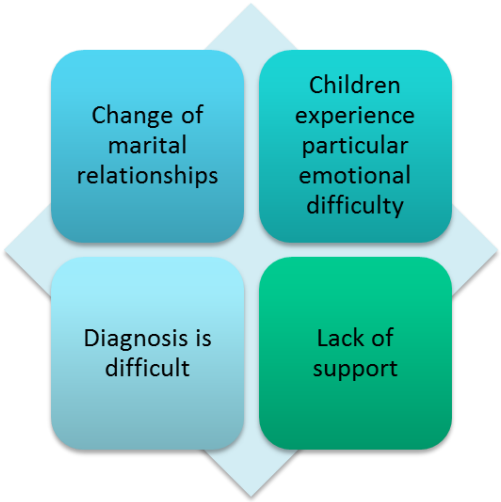
Challenges of YOD

Impact on patient and family

YOD has a profound impact on marital and other social relationships, often involving children. If the person has children it is important that they receive appropriate support and learn to understand what dementia is, how it affects their parent, what changes they may expect, and how to deal with them. Problematic behaviours are more frequent than in late-onset dementia, particularly the behavioural changes that are associated with Frontotemporal dementia, such as disinhibition, loss of empathy and inappropriate social conduct.

Typically YOD leads to premature retirement and reduced family income. Since people with YOD may work, engage in voluntary activities and have household resposibilities, financial and legal matters need attention. It is also important to plan for the future when the person may not have the capacity to deal with financial and legal issues.

Due to better general health and fewer physical co-morbid conditions, people with YOD spend longer living with the disease than individuals with late-onset dementia. Therefore care and support may be required for a longer period of time, and the cost of illness is likely to be higher.



Diagnosis may be difficult

The rarity of YOD, along with the wide spectrum of symptoms makes prompt diagnosis difficult. Some types of dementia (Frontotemporal degenerations) which affect the frontal lobes of the brain, are associated with profound alterations in personality and social conduct. Since impairment of language or vision may be the most prominent features of YOD (rather than decline in memory), obtaining a timely and accurate diagnosis may be especially difficult. The range of underlying causes also includes neurological disorders that lead to movement abnormalities and may require specific treatment and care. Furthermore, genetic causes play a far greater role in YOD than in late-onset dementia. Therefore, genetic counselling and genetic testing are important issues that require the involvement of additional specialists.

Lack of specific support and services

Existing health and social care structures, including day centres and special dementia services, often do not meet the needs of this group of patients in any country. Day centres and special dementia care units in nursing homes designed for the elderly may be inappropriate for younger individuals. Specific services for people with YOD, such as support groups, educational courses or leisure activities, have only recently been introduced and they are still rare. Cognitive, physical and behavioural training programmes remain an exception. Specific pharmacological treatment is currently unavailable for most people with YOD. Therefore, counselling and support for family and other carers are an extremely important component of ongoing care.

Where to get help?

Alzheimer associations

National and local Alzheimer's associations can provide valuable information on counselling, support and services that are available in an area. These organisations will also be able to answer specific questions related to living with dementia. Help will be provided with finding a healthcare professional with expertise in diagnosing and treating YOD; organising care; dealing with legal issues; and identifying an appropriate day centre, patient or carer support group, or nursing home.

Addresses of the national Alzheimer Associations can be found on the websites of

- **Alzheimer Europe:** http://alzheimer-europe.org/Alzheimer-Europe/Who-we-are/Our-members and
- **Alzheimer's Disease International:** http://www.alz.co.uk/finding-help

In the United Kingdom you can get additional support from the charity

- **YoungDementia UK:** www.youngdementiauk.org

Memory clinics

Memory clinics are specialised clinics that focus on memory disorders as well as other neurological disorders. Some are connected to universities and major medical centers; others are located within old age services or dementia research centres; and some are privately run. Memory clinics can provide services relating to the diagnosis and treatment of dementia, including cognitive training and medication management.

RHAPSODY—Research to Assess Policies and Strategies for Dementia in the Young
The main goal of RHAPSODY is to improve care for people with YOD by supporting their carers. The project will develop a web-based, interactive learning programme which is tailored to the needs of this particular group of people.

A multi-disciplinary consortium of eight partner institutions from six countries – France, Germany, the Netherlands, Portugal, Sweden and the United Kingdom are joining their efforts. The research teams represent academia, industry as well as a patient and carer advocacy organization.

RHAPSODY is an **EU Joint Programme Neurodegenerative Disease Research (JPND) project**. The project is supported through the following funding organisations under the aegis of JPND: France, The French National Research Agency (ANR); Germany, Federal Ministry of Education and Research (BMBF, FKZ: 01ED1404B); Netherlands, The Netherlands Organization for Health Research and Development (ZonMW); Portugal, Fundação para a Ciência e a Technologia (FCT); Sweden, Swedish Research Council (SRC); United Kingdom, Economic and Social Research Council (ESRC).

JPND is the largest global research inititiave aimed at tackling the challenge of neurodegenerative diseases. JPND aims to increase coordinated investment between participating countries in research aimed at finding causes, developing cures, and identifying appropriate ways to care for those with neurodegenerative diseases. www.jpnd.eu

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