

## *Dementia: a bibliographic refresh*

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### ABSTRACT

The last 50 years life expectancy at birth increased by almost 10 years, on average, across European Union. Dementia is not an inevitable part of aging. However, as population ages, the number of people living with dementia is set to increase as mainly affects older people, though it can start before the age of 65. Dementia is a syndrome due to disease of the brain, usually of a chronic or progressive nature, in which there is disturbance of multiple higher cortical functions, including memory, thinking, orientation, comprehension and calculation. It's an umbrella term that covers a number of specific illnesses like Alzheimer's, Vascular Dementia, Parkinson's, Huntington's, Lewy body disease, Creutzfeldt-Jacob, Korsakoff's and other related dementias. Unfortunately, many people cannot recognize these symptoms showing that something is wrong or mistakenly assume that these symptoms are a normal part of the aging process. Other times they refuse to react even when they know that something is wrong. All health care professionals are faced with these wrong perceptions and nobody can avoid them. In addition, the stigma of being elderly and suffering from mental illness increases alienation and social exclusion. A limited future and a frozen past is the situation that these patients face daily. Over the last few years, researchers have focused on the development of new drugs and treatments to prevent or reverse the disease. Improved co-operation and co-ordination of healthcare professionals is important to make progress in the care of these patients and mental health nurses are an important part of it.



**Keywords:** Dementia, Alzheimer's, Parkinson's, Huntington's, Creutzfeldt-Jacob



Citation

A. Douchty, C. Staikos, D. Louloudis. Dementia: a bibliographic refresh. *Scientific Chronicles* 2018; 23(3): 303-315

eoi: <http://eoi.citefactor.org/10.11212/exronika/2018.3.5>

## INTRODUCTION

The last 50 years, life expectancy at birth increased by almost 10 years, on average, across European Union. This is due largely to improved socio-economic and environmental conditions, as well as the best medical treatment and intensive care. On average, a European born in 2014 is expected to live 80.9 years. In most regions of European Union, the relative percentage of elderly population is increased progressively, as a result of the significant and continuous increase in life expectancy, as well as the retirement of the generation of demographic explosion afterwards World War II. Since then, the birth rate has decreased progressively, and life expectancy has increased gradually, resulting in a slowdown in the natural population growth rate. Until 2003, this figure has been almost balanced, although this course has generally been reversed with the onset of the financial and economic crisis: between 2008 and 2013, but it has risen again in 2014. This fact involves that today about 18.9% of the population consists of elderly people (people aged 65 years and older). [1]

Dementia is not an inevitable part of aging. However, as population ages, the number of people living with dementia is set to increase as mainly affects older people, though it can start before the age of 65. There are about 676,000 people with dementia in England and this number is expected to grow in the next 30 years. [2]

## DEFINITION

According to International Statistical Classification of Diseases and Related Health Problems (ICD), Dementia is a syndrome due to disease of the brain, usually of a chronic or progressive nature, in which there is disturbance of multiple higher cortical functions, including memory, thinking, orientation, comprehension, calculation. This syndrome occurs in Alzheimer's disease, in cerebrovascular disease, learning capacity, language, and judgement. Consciousness is not clouded. The impairments of cognitive function are commonly accompanied, and occasionally preceded, by deterioration in emotional control, social behavior, and motivation, and in other conditions primarily or secondarily affecting the brain.

In the Fifth Edition (2013) of Diagnostic and Statistical Manual of Mental Disorders (DSM-V), dementia is called Major Neurocognitive Disorder (NCD) but the term dementia will still be often used. [3]

## TYPES OF DEMENTIA

### 1. Dementia in Alzheimer's disease.

Named after Dr. Alois Alzheimer. In 1906, Dr. Alzheimer noticed changes in the brain tissue of a woman who had died of an unusual mental illness. Her symptoms included memory loss, language problems and unpredictable behavior. After her death, he examined her brain and found many abnormal clumps (now called amyloid plaques) and tangled bundles of fibers (now

called neurofibrillary, or tau, tangles. Another feature is the loss of connections between nerve cells (neurons) in the brain. Neurons transmit messages between different parts of the brain, and from the brain to muscles and organs in the body. Is currently ranked as the sixth leading cause of death in the United States, but recent estimates indicate that the disorder may rank third, just behind heart disease and cancer, as a cause of death in older people [4]. Alzheimer's disease is the most common type of dementia, affecting an estimated 850,000 people, in the UK also. The symptoms of Alzheimer's disease progress slowly over several years. Quite often these symptoms are confused with other conditions and may initially be put down to old age. The rate at which the symptoms progress is different for each individual and it's not possible to predict exactly how quickly it will get worse.

Generally, the symptoms of Alzheimer's disease are divided into three main stages.

a) **Early symptoms:** In the early stages, the main symptom of Alzheimer's disease is memory lapses. For example, someone with early Alzheimer's disease may:

- forget about recent conversations or events, or misplace items
- forget the names of places and objects, or have trouble thinking of the right word
- repeat themselves regularly, such as asking the same question several times
- show poor judgment or find it harder to make decisions
- become less flexible and more hesitant to try new things

There are often signs of mood changes, such as increasing anxiety or agitation and periods of confusion.

b) **Middle-stage symptoms:** As Alzheimer's disease develops, memory problems will get worse. Someone with this condition may find it increasingly difficult to remember the names of people they know and may struggle to recognize their family and friends. Other symptoms may also develop, such as:

- increasing confusion and disorientation – for example, getting lost, or wandering and not knowing what time of day it is
- obsessive, repetitive or impulsive behavior
- delusions (believing things that are untrue) or feeling paranoid and suspicious about cares or family members
- problems with speech or language (aphasia)
- disturbed sleep
- changes in mood, such as frequent mood swings, depression and feeling increasingly anxious, frustrated or agitated
- difficulty performing spatial tasks, such as judging distances
- hallucinations

At this stage, someone with Alzheimer's disease usually needs support to help them with their everyday living. For example, they may need help eating, washing, getting dressed and using the toilet.

c) **Later symptoms:** In the later stages of Alzheimer's disease, the symptoms become increasingly severe and distressing for the person with this condition, as well as their carers, friends and family. Hallucinations and delusions may come and go over the course

of the illness but can get worse as the condition progresses. Sometimes people with Alzheimer's disease can be violent, demanding and suspicious of those around them. Several other symptoms may also develop as Alzheimer's disease progresses, such as:

- difficulty eating and swallowing (dysphagia)
- difficulty changing position or moving around without assistance
- considerable weight loss – although some people eat too much and put on weight
- unintentional passing of urine (urinary incontinence) or stools (bowel incontinence)
- gradual loss of speech
- significant problems with short- and long-term memory

In the severe stages of Alzheimer's disease, people may also need full-time care and assistance with eating, moving and using the toilet. [5]

## **2. Vascular dementia.**

This type of dementia is a decline in thinking skills caused by conditions that block or reduce blood flow to various regions of the brain, depriving them of oxygen and nutrients. Inadequate blood flow may damage and eventually kill cells anywhere in the body, but the brain is especially vulnerable. Changes in thinking skills sometimes occur suddenly after a stroke, which blocks major blood vessels in the brain. Thinking difficulties may also begin as mild changes that gradually worsen as a result of

multiple minor strokes or another condition that affects smaller blood vessels, leading to widespread damage. Vascular brain changes often coexist with changes linked to other types of dementia, including Alzheimer's disease and dementia with Lewy bodies. Several studies have found that vascular changes and other brain abnormalities may interact in ways that increase the likelihood of dementia diagnosis. [6] Vascular dementia affects people in different ways and the symptoms depend on the areas of the brain that have been damaged.

Initially, patients present as follows:

- Not being able to understand or respond to things very quickly
- Becoming confused
- Not being able to remember things
- Finding it difficult to concentrate
- Not being able to find the right word when speaking
- Seeming down or depressed

As the disease progresses patients present as follows:

- Behaving in a different way, being aggressive or behaving inappropriately and not being able to control emotions
- Finding it difficult to walk or keep balance
- Having problems controlling bladder
- Seeing or hearing things that aren't there (hallucinations)
- Believing things that aren't true (delusions)

Sometimes symptoms of vascular dementia can be confused with the effects of stroke. The main difference is that vascular dementia gets worse over time [7]. After a

large or medium-sized blood vessel is blocked by a clot the stroke may be so small that the person doesn't notice any symptoms. Alternatively, the symptoms may only be temporary - lasting perhaps a few minutes - because the blockage clears itself. If symptoms last for less than 24 hours this is known as a 'mini-stroke' or Transient Ischaemic Attack (TIA). If the blood supply is interrupted for more than a few minutes, the stroke will lead to the death of a small area of tissue in the brain. This specific area is known as an infarct. [8]

a) **Vascular dementia of acute onset** (ICD-10) or single infarct dementia caused when one infarct forms by cerebrovascular thrombosis, embolism or hemorrhage, in a significant part of the brain. It is characterized by the sudden onset of changes in thinking skills of behavior after a stroke. The symptoms depend on the location of the stroke and what brain functions are affected by the damage. Provided no further strokes occur, the person's symptoms may remain stable or even get better over time. However, if there is other vascular disease also affecting the brain or additional stroke occurs, symptoms may get worse.

b) **Multi-infarct dementia** is caused by multiple strokes, which produce an accumulation of infarcts in the cerebral parenchyma. Reasoning and thinking skills may be affected to the point that a diagnosis of vascular dementia is made. Other

symptoms may include depression and mood swings, but the symptoms very much depend on the location of the brain damage and can have a step-wise progression. After a new stroke, symptoms get worse and then are stabilized for a time. [9]

c) **Binswanger's disease (BD)**, also called subcortical vascular dementia, is a type of dementia caused by widespread, microscopic areas of damage to the deep layers of white matter in the brain. The damage is the result of the thickening and narrowing (atherosclerosis) of arteries that feed the subcortical areas of the brain. Atherosclerosis (commonly known as "hardening of the arteries") is a systemic process that mainly affects blood vessels throughout the body. It begins late in the fourth decade of life and increases in severity by age. As the arteries become more and more narrowed, the blood supplied by those arteries decreases and brain tissue finally dies. The symptoms associated with BD are related to the disruption of subcortical neural circuits that control what neuroscientists call executive cognitive functioning: short-term memory, organization, mood, the regulation of attention, the ability to act or make decisions and appropriate behavior. The most characteristic feature of BD is psychomotor slowness - an increase in the length of time it takes, for example, for the fingers to turn the thought of a letter into the shape of a letter on a piece of paper. Other symptoms include forgetfulness (but not as severe as the forgetfulness of Alzheimer's disease), changes in speech, an unsteady gait, clumsiness or

frequent falls, changes in personality and mood (most likely in the form of apathy, irritability, and depression), and urinary symptoms that aren't caused by urological disease. Brain imaging, which reveals the characteristic brain lesions of BD, is essential for a positive diagnosis. [10]

### 3. Mixed cortical and subcortical vascular dementia.

This is a condition in which abnormalities characteristic of more than one type of dementia occur simultaneously in the brain. Physicians may also name this condition "Dementia - multifactorial." In the most common form of mixed dementia, the abnormal protein deposits associated with Alzheimer's disease coexist with blood vessel problems linked to vascular dementia. In some cases, a person may have brain changes linked to Alzheimer's disease, vascular dementia and dementia with Lewy bodies. [11]

### 4. Lewy Body Dementia

This is a disease related with abnormal deposits of a protein called alpha-synuclein in the brain. These deposits, called Lewy bodies and affect a few different brain regions like:

- the cerebral cortex, that controls many functions, including information processing, perception, thought, and language
- the limbic cortex, which plays a major role in emotions and behavior

- the hippocampus, which is essential to forming new memories
- the midbrain, including the substantia nigra, which is involved in movement
- areas of the brain stem important in regulating sleep and maintaining alertness
- brain regions important in recognizing smells (olfactory pathways)

Lewy body dementia includes two related conditions, dementia with **Lewy bodies** and **Parkinson's disease** dementia. The difference between them lies largely in the timing of cognitive (thinking) and movement symptoms. In dementia with Lewy bodies, cognitive symptoms are noted within a year of parkinsonism, any condition which involves the types of movement changes seen in Parkinson's disease. In Parkinson's disease dementia, movement symptoms are most pronounced, with cognitive symptoms developing years later. [12]

a) **Parkinson's Disease** is the most common disease after Alzheimer's. There are more than 1.2 million people living with Parkinson's in Europe. The average age of onset is 60 years, although more than one in ten people are diagnosed before the early age of 50. The condition was first described by Dr James Parkinson (1817) in which he reported in detail the symptoms. In the 1960s it was discovered that the symptoms are primarily related to a lack of a neurotransmitter (dopamine) as a result of degeneration of dopamine producing neurons within the

substantia nigra in the basal ganglia in the mid-brain. [13] The symptoms of Parkinson's disease usually develop gradually and are mild at first.

Motor Symptoms that impact physical movement are:

- **Tremor** is an uncontrollable movement which affects a part of the body.
- **Essential Tremor**, the resting tremor of Parkinson's is an instinctive reflex of the hands, head, legs, body or voice and is more likely to occur when the limb is relaxed and resting.
- **Dystonic tremor** is a range of movement disorders that cause painful muscle spasms and contractions. [14]
- **Bradykinesia** (slowness of movement) describes the slowness in performing physical movements and affects walking, speech, swallowing and speaking.
- **Rigidity** means stiff or inflexible muscles, preventing muscles from stretching and relaxing as they should, reducing facial expression (mask-like face) and painful muscle cramps.
- **Postural Instability** (poor balance).

**Parkinsonism** is a generic term for the clinical syndrome involving the above main motor symptoms that can be seen in someone with Parkinson's disease. Other motor symptoms are:

- **Dyskinesia** often happen in several medical conditions and is the term used to describe unintentional, nonvoluntary and uncontrollable movements. These include twitches, jerking, twisting or simple restlessness but not tremor.
- **Freezing of gait**, the inability to start or continue walking, characterized by difficulty in stepping forward (at initiation or during

walking), and inability to lift the foot from the floor. [15]

- **Falls**
- **Restless Legs Syndrome (RLS)**, also called Willis-Ekbom disease (WED), is a common movement disorder characterized by an irresistible urge to move the legs. The sensations in patients' legs are often difficult to define but may be described as aching throbbing, pulling, itching, crawling, or creeping. These symptoms tend to occur most during quiet wakefulness, for example when watching a movie or during sleep. This is known as periodic limb movement during sleep (PLMS) and periodic limb movement while awake (PLMW). [16]
- **Motor fluctuations** occurs when the benefit from levodopa wearing off before the next dose is due, usually predictable or when rapid and unpredictable fluctuations between 'on' and 'off' periods, due to fluctuating responses to levodopa, usually after several years of use. Also occurs when there is a failure of anti-parkinsonian medication to provide symptom relief.

Mental health problems include:

- Dementia
- Anxiety
- Depression
- Hallucinations and delusions
- Dopamine Dysregulation Syndrome, characterised by both addictive and stereotyped behaviour. It is defined as compulsive overuse or addiction in dopaminergic drugs, usually associated with punding, pathological gambling, eating, shopping or hypersexuality. [17]

#### Autonomic Dysfunction:

- Bladder problems including frequency and incontinence
- Constipation
- Sialorrhea, excessive production of saliva (drooling)
- Hyperhidrosis, excessive sweating and increased sensitivity to temperatures
- Anosmia refers to a decrease or loss of sense of smell.
- Dysphagia, swallowing difficulties and weight loss
- Orthostatic or postural hypotension
- Nerve pain that can cause unpleasant sensations, such as burning, coldness or numbness
- Sexual dysfunction

#### Other symptoms:

- Sleep disturbance and daytime hypersomnolence.
- Aspiration pneumonia
- Fatigue it is typically described as extreme tiredness, exhaustion or a complete lack of energy which limits what you can do and affects quality of life. [18]

b) **Dementia with Lewy bodies** initially appears as a decline in cognitive skills which resembles Alzheimer's disease. Over time, however, distinctive symptoms develop which suggest DLB. These may include:

- visual hallucinations
- fluctuations in cognitive ability, attention and alertness;
- slowed movement, difficulty walking or rigidity (parkinsonism);
- sensitivity to medications used to treat hallucinations;

- REM sleep behaviour disorder in which people physically act out their dreams;
- more difficulty with executive function than memory; executive function means performing complex mental activities such as multi-tasking, problem solving, spatial awareness and analytical thinking. [19]

#### 5. Dementia in other diseases classified elsewhere.

Cases of dementia to causes other than Alzheimer's disease or cerebrovascular disease.

a) **Frontotemporal lobar degeneration or dementia (FTD) or Pick's disease (PiD)** was named after Arnold Pick, a German neurologist, who first described this rare neurodegenerative disease in 1892. Clinically, there are two core PiD symptomatic patterns involving behavior and language. Features of behavioral changes can be either impulsive (disinhibited) or bored (apathetic) and include inappropriate social behavior; lack of social tact or empathy, loss of insight into the behaviors of oneself and others, an increased interest in sex, changes in food preferences, agitation or, conversely, blunted emotions, neglect of personal hygiene; repetitive or compulsive behavior, and decreased energy and motivation. Features of language disturbance include difficulty in thinking or understanding speech, often in conjunction with abnormal behavioral symptoms but with intact spatial skills and memory. Extrapyramidal features such as rigidity, gait instability, a masked face and micrographia are frequently encountered but a resting tremor is rare.

Fasciculation, muscle wasting, motor weakness, swallowing difficulty with an attenuated gag reflex and effortful speech may be seen. [20]

**b) Creutzfeldt-Jakob Disease (CJD) is a rare, fatal brain disease.** It is caused by an abnormal form of a substance called the prion protein. In its normal form, prion protein is made by most body cells and doesn't cause disease. In its abnormal form, prion protein is toxic to brain cells and causes disease. The brain damage in people or animals with prion disease may be seen with a microscope. One kind of damage is "spongiform change", in which the brain tissue looks like a sponge with many tiny holes. Many of the brains nerve cells die.

Most cases (about 90%) occur in older people, without warning or a clear reason. This is called sporadic CJD, meaning it happens occasionally or unpredictably and begins with the forming of abnormal prion by one or a few brain cells and then spreads to the normal prion in the rest of the brain.

Most other CJD cases (about 10%) have genetic changes called mutations. These are found in the gene that tells body cells how to make prion protein and increase the chances that the prion protein will become abnormal and that prion disease will develop. Infection with human prions has occurred by accident through certain medical procedures involving human tissues. This kind of CJD is called iatrogenic, meaning it is caused by a medical treatment.

Some cases have resulted from human exposure to Bovine Spongiform Encephalopathy (BSE, commonly referred to as "mad cow disease"), a prion disease of cattle. These patients developed a new form called variant CJD.

The symptoms of CJD may vary greatly from person to person. Often a mental or neurological problem appears first. However, early symptoms may seem mild, sometimes like depression. A family member is often the first to notice mood swings, social withdrawal or lack of interest. After it starts, CJD usually progresses rapidly. Eventually the person loses the ability to move, speak or care for themselves, and needs full-time care.

Most people die within 6 months from when the illness begins. Some can live as long as 1 year, but rarely longer. The symptoms are similar to sporadic and iatrogenic CJD and include:

- dementia (loss of memory and thinking abilities)
- ataxia (unsteadiness when walking or standing, marked clumsiness)
- myoclonus (sudden jerky movements)
- psychological problems (depression, irritability, changes in behavior)
- vision problems (including blindness)
- aphasia (loss of ability to speak or understand speech)
- stiffness of arms or legs
- difficulty swallowing

More than 50 different rare genetic mutations have been found in the prion protein gene. Individuals carrying different

mutations usually have different symptoms. Symptoms can differ enough that some genetic prion diseases have been given special names, such as:

- Genetic CJD, symptoms are similar to those of sporadic CJD
- Gerstmann-Sträussler-Scheinker disease (GSS), usually starts with a clumsiness or unsteadiness when standing or walking, and later progresses to dementia. The disease usually lasts longer than sporadic or genetic, and the person may survive for several years after the illness begin.
- Fatal familial insomnia (FFI) the main symptom is a severe, progressive and untreatable form of insomnia. FFI leads to reduce control of basic bodily functions, such as blood pressure. Coma and death eventually follow. [21]

**c) Huntington's disease (HD) or Huntington's Chorea (HC)** is an inherited disorder of the central nervous system (each child has a 50% risk of developing the disease) and caused by a faulty gene on chromosome 4, which produces a protein called Huntingtin. This faulty gene leads to damage of the nerve cells in areas of the brain including the basal ganglia and cerebral cortex. HD usually develops in adulthood and affects both men and women. [22]

Unfortunately, many people cannot recognize these symptoms showing that something is wrong or mistakenly assume

that these symptoms are a normal part of the aging process. Other times they refuse to react even when they know that something is wrong. All health care professionals are faced with these wrong perceptions and nobody can avoid them. In addition, the stigma of being elderly and suffering from mental illness increases alienation and social exclusion. A limited future and a frozen past are the situations that these patients face daily. Over the last few years, researchers have focused on the development of new drugs and treatments to prevent or reverse the disease.

The Psycho-geriatric patients still are the main users of social and health care services. The increased discharge rate of these patients creates serious demands on Community care systems, and it is absurd to nursing practice to focus more in the hospital environment, which maintains the borderline between hospital and community care. This creates the need for a role that will act as a link between hospitals and community services, the role of community psychiatric nurse, a role that already exists in several European countries. Improved co-operation and co-ordination of healthcare professionals is important to make progress in the care of these patients and nurses are an important part of it.

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## ΑΝΑΣΚΟΠΗΣΗ

### *Άνοια: μια βιβλιογραφική ανασκόπηση*

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#### ΠΕΡΙΛΗΨΗ

Τα τελευταία 50 χρόνια το προσδόκιμο ζωής αυξήθηκε κατά σχεδόν 10 χρόνια, κατά μέσο όρο, σε ολόκληρη την Ευρωπαϊκή Ένωση. Η Άνοια δεν αποτελεί αναπόφευκτο μέρος της γήρανσης, ωστόσο καθώς ο πληθυσμός γερνάει, ο αριθμός των ατόμων που ζουν με άνοια αυξάνεται καθώς επηρεάζει κυρίως τους ηλικιωμένους, αν και μπορεί να ξεκινήσει και πριν από την ηλικία των 65 ετών..Η άνοια είναι ένα σύνδρομο που οφείλεται σε νόσους του εγκεφάλου, συνήθως χρόνιας ή προοδευτικής φύσης, όπου υπάρχει διαταραχή πολλαπλών ανώτερων λειτουργιών του φλοιού του, συμπεριλαμβανομένων της μνήμης, σκέψης, προσανατολισμού, κατανόησης και του υπολογισμού. Είναι ένας όρος ομπρέλα που καλύπτει μια σειρά συγκεκριμένων ασθενειών όπως

Alzheimer's, Αγγειακή Άνοια, Parkinson's, Huntington's, Lewy Body, Creutzfeldt-Jacob, Korsakoff's και άλλες σχετιζόμενες με την Άνοια νόσους. Δυστυχώς, πολλοί άνθρωποι δεν μπορούν να αναγνωρίσουν αυτά τα συμπτώματα δείχνοντας ότι κάτι είναι λάθος ή λανθασμένα υποθέτουν ότι αυτά τα συμπτώματα είναι φυσιολογικό μέρος της διαδικασίας γήρανσης. Άλλες φορές αρνούνται να αντιδράσουν ακόμη και όταν ξέρουν ότι κάτι δεν πάει καλά. Όλοι οι επαγγελματίες υγείας αντιμετωπίζουν αυτές τις λανθασμένες αντιλήψεις και κανείς δεν μπορεί να τις αποφύγει. Επιπλέον, το Στίγμα ότι είναι ηλικιωμένοι και πάσχουν από ψυχικές ασθένειες αυξάνει την αποξένωση και τον κοινωνικό αποκλεισμό. Ένα περιορισμένο μέλλον και ένα παγωμένο παρελθόν είναι η κατάσταση που αντιμετωπίζουν αυτοί οι ασθενείς καθημερινά. Τα τελευταία χρόνια, οι ερευνητές επικεντρώθηκαν στην ανάπτυξη νέων φαρμάκων και θεραπειών για την πρόληψη ή την αναστροφή της νόσου. Η βελτίωση της συνεργασίας και του συντονισμού των επαγγελματιών στον τομέα της υγείας είναι σημαντική για την πρόοδο στη φροντίδα αυτών των ασθενών και οι νοσηλευτές Ψυχικής Υγείας αποτελούν σημαντικό μέρος αυτής.



**Λέξεις ευρετηρίου:** Άνοια, Alzheimer's, Parkinson's, Huntington's, Creutzfeldt-Jacob



Παραπομπή

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