ALZHEIMER’S DISEASE AND RELATED ILLNESSES

HELPING YOU TO GET THE MOST OUT OF LIFE
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An important note for English speaking readers: in France the term dementia is considered pejorative and to some shocking. These days English speakers use the term freely without any perceived problems. One might say that using the term dementia in France is akin to saying someone is a lunatic in English. So, when in this document the terms “la Maladie Alzheimer et les Maladies Apparentées” or translated “Alzheimer’s disease and related diseases”, this means dementia. Alzheimer’s disease is one of a number of types of dementia as are, for instance, Vascular Dementia and Dementia with Lewy Bodies. These other forms of dementia are also described later in this booklet in the section “Related Illnesses.”
INTRODUCTION

Alzheimer’s disease is a “neurodegenerative” brain disorder leading to the progressive disappearance of neurons.

It causes cognitive changes which affect memory, language and reasoning etc. The spread of brain damage causes other problems which gradually erode the independence of the person affected.

The illness appears most often in older people, but it is not a normal consequence of ageing.

SYMPTOMS

What are the first signs of the disease?

The illness initially presents through memory problems but...

... we need to distinguish between harmless forgetfulness and abnormal loss of memory. Forgetting an appointment, a code number or where you put your keys, can happen to anyone and may be due to a lack of attention. It is also not unusual, on occasion, to have difficulty remembering the names of people we know.

With Alzheimer’s disease memory disorders can take an unusual turn; the person affected may forget important family events.

Problems with memory are not the only signs of the disease. Odd changes in daily behaviour (especially in money management, travelling, using the telephone or taking medication), the inability to find words, confusion relating to place and time, difficulties following a conversation, changes in behaviour: these are all clues which may alert family and friends.

However, this is a gradual shift and may not be noticed by the family circle, especially when the person “compensates” for their difficulties.
Is it right to talk about a “disease of the memory”?

In a sense yes, because the first area of the brain affected by the illness is the hippocampus, a brain structure essential for memorising new information.

Generally, the person affected finds it increasingly difficult to remember “new information. They cannot recall what they have just done or keep asking the same questions every few minutes. The ability to create new memories is altered, but conversely, distant memories, such as school or their first job are preserved.

Over the months and years, as brain damage spreads, the person will have trouble recalling older memories. In fact, this inability to recover memories “recedes” through the person’s life, from the most recent events to those in the distant past.

In the long run, memory dysfunction brings about a different perception of reality. Although the person has “forgotten” twenty years of memories, at times they believe they are living their daily life of twenty years ago.

However, Alzheimer’s disease cannot simply be summed up by memory problems. All the cognitive faculties, formerly called intellectual functions, are affected.

Alzheimer is talked about as the “disease of the 4 As” – amnesia, apraxia, agnosia, aphasia - to which we should add problems with orientation and with executive function.

What is aphasia?

This is characterised by language trouble. The person has difficulty speaking and/or understanding what they are told.

At first, these difficulties only involve a few words, usually the most complicated. Then even simple words cause problems. The person affected uses inappropriate words, invents words or keeps repeating the same expression. These problems may develop until the person lapses into silence.

Language problems occur in the great majority of cases, although in many people they are not too noticeable.

What is apraxia?

This is characterised by problems with movement or actions.

At the onset of the illness these problems can be detected in a poorer quality of writing and drawing. Then difficulties arise with more complex activities requiring some skill, such as knitting, DIY or cooking.

Problems with movement lead to considerable loss of independence, requiring the involvement of a third person. For help with intimate matters, it is better to engage a professional, in order not to upset the relationship between the person affected with Alzheimer’s and the family carer.

What is agnosia?

This is characterised by problems with recognition. Sensory perception is intact, but the person affected can no longer identify objects and/or people.

2 very specific types of agnosia

Failure to recognise faces, often wrongly attributed to memory problems. Many people affected compensate by the sound of the voice.

Failure to acknowledge the disease. The person affected is not aware of their problems and does not think they are ill. This agnosia is sometimes confused with denial, which is a psychological defence mechanism.
What are the problems of orientation?
There are two types:

• **Temporal disorientation**
The inability to place oneself in time. The problem develops in a “decreasing” pattern: at first, the person affected does not know what year it is, then they don’t know the season, month, day of the week or time etc.

• **Spatial disorientation**
The inability to place oneself in spatial surroundings. The problem will first be experienced in unknown places, then the person is unable to place themselves in their neighbourhood, house, bedroom etc.

What characterises problems of “executive function”?
A change in the abilities needed to carry out an action:

• Concentration, or the ability to focus on a task.
• Reasoning, including the ability to understand the process of a mechanism and conceptualise it.
• Planning, or the ability to conceive and carry out, in a logical order, the steps making up an action.

Is depression a sign of the illness?
The feeling of losing your independence and abilities naturally brings with it a lowering of morale which can lead to depression. In this case, depression is a consequence of the illness, which frequently occurs before a diagnosis and recurs afterwards.

Doctors also wonder whether depression could be a “risk factor”, something which might contribute to the onset of the illness. This however is only a hypothesis.

The fact remains that most cases of depression in the elderly have nothing to do with Alzheimer’s disease. These should be diagnosed and treated as such.

Why behavioural problems?
Behavioural problems, also called Psychological and Behavioural Symptoms of Dementia (PBSB) are changes in the behaviour of the affected person which appear after the onset of the illness or following an earlier assessment. These symptoms occur frequently and tend to get worse as the illness develops.

Brain dysfunction alone is not solely responsible for behavioural problems. Other factors such as environment, life history, attitudes of friends and family or a physical problem can have an influence.

These problems often express a need. The carer must try to identify this need so they can provide an appropriate response.

There are not always grounds for prescribing drug treatments. The effectiveness of psychotropic drugs has been questioned, and their prescription should remain careful and controlled.

An example of trouble with executive function
Drinking a glass of water involves planning several steps: taking the bottle, undoing the cap, pouring the water into the glass, bringing it to the mouth.

An example of behavioural problems
The patient wanders because they are disoriented or attacks their carer because they no longer recognise that person.
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ORIGINS AND PROGRESSION

What is the characteristic damage caused by the disease?
Alzheimer's disease is the result of two types of damage to the central nervous system.

- **Neuro-fibrillar or “tangle” degeneration** The occurrence of Tau protein abnormalities inside the nerve cells.

- **Amyloid or “senile” plaques** These are deposits of the amyloid Beta protein outside nerve cells. The damage gradually spreads to different areas of the cerebral cortex, where it remains unnoticed for a long time. The plaques then lead to visible symptoms as they gradually affect areas important for brain function.

Is Alzheimer's disease an “elderly person’s” illness?
Until 1960-1970, intellectual decline was considered a natural consequence of ageing. It was normal to become “spoiled” or “return to childhood”. This received wisdom, unfortunately, still pervades some thinking today. Although the risk of contracting the disease increases with age, the processes of Alzheimer's disease and that of normal ageing are very different.

An unaffected elderly person carries out tasks in the same way as a young person but needs more time and effort. A person with Alzheimer's disease however, is no longer able to perform certain tasks.

Do we know what causes the disease?
Although researchers have shed light on the processes that lead to brain damage, they have not yet identified what causes them. However, they have discovered factors which may contribute to the development of the disease, or so-called risk factors.

Among these risk factors are:

- **Age**
  - older people are most affected

- **Tobacco**

- **Physical inactivity**

- **Low level of education**

- **Obesity**

- **Diabetes**

- **Hypertension**

The relative effects of each of these risks have yet to be evaluated in long-term studies.

Is Alzheimer’s disease hereditary?
There are so-called “familial” forms of Alzheimer’s disease which are linked to mutations of certain genes. There are very few of these and they only involve around 1% of all people affected by the disease.

In these variants, relatives carrying the mutation often trigger the disease early, sometimes before the age of forty. In the families concerned, large numbers of people are affected by the illness.

In the other cases (99%), the disease is labelled “sporadic”. Its onset is caused by many factors, often resulting from the combination of an existing predisposition and other risk factors.

The impact of this predisposition varies with each individual case and appears to be linked to specific genetic profiles. It has been shown that the effect of apolipoprotein E, which exists in three forms, depends on each individual, carriers of type 4 being more likely to develop the disease, while type 2 plays a protective role.

How many people are affected?
In 2008, according to the Health Department, 400 000 people over 60 were reported as suffering from a “long term illness” or being treated for Alzheimer’s disease or related illness. However, epidemiology studies show that almost half of people affected do not have a diagnosis. If they did, the number of French people affected by Alzheimer’s disease or a related illness would exceed the million mark.

At what age does Alzheimer’s disease appear?
The disease mainly affects people over the age of 65, but according to estimates, about 32 000 French people under 65 are affected. Some very rare cases are also detected before the age of 40. Some very rare cases are also detected before the age of 40.
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This susceptibility for predisposition explains why, when several cases occur within a family, the risk of contracting the disease is increased, although this is not inevitable. Taking action to mitigate other risk factors (high blood pressure, diabetes) then becomes extremely important.

Other preventive measures are also vital, especially where diet (one rich in fruit and vegetables and restricting animal fat intake) and intellectual and physical activities are concerned.

What are the stages of the disease?
Not everyone is affected at the same pace and in the same way. Nevertheless, several stages can be defined:

- **The pre-clinical phase**
  Damage caused by the disease is present but does not show.

- **The early (mild) phase**
  Short term memory deteriorates. The patient clearly remembers childhood memories but cannot recall what they just said on the telephone. These changes are often wrongly considered normal signs of ageing. There may also be difficulties in expressing oneself, lowness of spirit or changes in mood (irritability, sadness).

- **The middle “moderate” phase**
  The severity of their symptoms impact on the person’s daily life, forcing them to leave their job if they are still working. Memory problems get worse and they begin to lose their independence. Personal care and dressing become difficult and life in the home is only possible with outside help. Some people find it increasingly difficult to express themselves verbally, others to keep track of time and space.

- **The advanced (severe) phase**
  The person affected is no longer able to eat, walk or go to the bathroom alone. They no longer really recognise members of their family and lose the use of language. Constant supervision is required and care in a specialised centre is often necessary.

  However, exchanges with the person affected can continue through non-verbal communication.

Does Alzheimer’s disease prevent people from living an active life?
No. Many people lead active and interesting lives for many years after a diagnosis. They have projects and plans, their lives always make sense. The typical general picture of the disease is often associated with the advanced or terminal stages of the illness, which is very simplistic. The pursuit of various activities in an appropriate setting and maintaining a social life can promote better health for the person affected.
What is frontotemporal degeneration?
In terms of pathology, it is the frontal and sometimes temporal areas of the brain which suffer damage. There are several types of frontotemporal degeneration:

• Pick’s disease and atypical frontal dementia
The symptoms are exactly the same. In Pick’s disease a post-mortem examination reveals the presence of distinctive cells, known as Pick bodies (after the Czech neurologist Arnold Pick). These cells do not occur in atypical frontal dementia. Note – today the term “Pick’s disease” is rarely used, referring instead to “frontotemporal dementia”.

• Semantic dementia
Initially, this is characterised by forgetting the meaning of words (objects, faces, etc) and concepts. The person complains about losing their vocabulary and no longer understands certain words. Speech is fluent but makes no sense. Later, the person presents character disorders, particularly selfishness and resistance to change or to accept new ideas. In the longer term, significant behavioural problems occur.

What should our attitude be towards the expression “dementia”?
In common parlance, the term dementia is associated with madness. Therefore, the use of this expression is not recommended, in order to avoid stigmatising those people affected. In strictly medical terms, dementia syndrome defines a developmental stage of Alzheimer’s and related diseases, meaning that changes in cognitive function have led to loss of independence in social and daily life.

Image 1: A woman and a child.

Neurodegenerative diseases can cause language difficulties (they can be some of the early symptoms) collectively called Aphasies Primaires Progressive or APP. These are not exclusive to any one of the different types of dementia, which since 2011 have been grouped in three classes.

The three groups are:
1 – APP - agrammatical
This is marked by grammatical mistakes, wrong choice of words (paraphasies) or by slow speech. This often signals the start of fronto-temporal degeneration (dégénérescence lobe fronto-temporal – DLFT), the progressive decay of neurones in the front area of the brain.

2 – APP – semantic
This is characterised by a loss of understanding of the meaning of words or a general understanding in speech. This can be seen in many forms of dementia although not DLFT. Here the person gives the impression of not hearing, but in fact this is because they do not understand what is being said.

3 – APP - logopenic
This is manifest when a person finds it difficult to find the right word to express themselves resulting in broken speech or use of words similar to those they intend such as using the word bowl instead of cup or boat instead of coat. This can be an early sign of Alzheimer’s disease.

What do we mean when we talk about neurodegenerative disease?
This disease is characterised by changes in essential neural structures, especially nerve cells (also called neurons) due to degenerative damage. The process is slow and gradual. 70% of dementias are neurodegenerative, the rest being non-degenerative or mixed.
What is Lewy body disease?
There are similar symptoms to Alzheimer’s disease with certain peculiarities, such as a wide variety of problems occurring in a single day, as well as difficulty walking (repeatedly falling backwards) and hallucinations, which are often visual but can also involve hearing or smell.

There is also a parkinsonian syndrome as well as sleep disorders with nightmares.

It is important to make an exact diagnosis so that antipsychotic drugs (neuroleptics) are not prescribed, as these can cause a marked deterioration of disorders and may be fatal. It should be noted that a diagnosis of dementia with Lewy bodies is often made in two stages because initially it can resemble Alzheimer’s or Parkinson’s disease.

What is vascular dementia?
Unlike Alzheimer’s disease, frontotemporal degeneration or Lewy body disease, this type of dementia is not degenerative.

Problems are caused by a stroke, due to a haemorrhage or embolism in the blood vessels of the brain.

Not all strokes lead to cognitive problems. We talk about vascular dementia if a patient presents with symptoms characteristic of dementia as the result of a stroke. It depends on the area of the brain affected and whether the remaining neurons are able to recover lost brain function.

It is possible to suffer a single major stroke with dramatic consequences: in this case, problems occur suddenly and brutally. But sometimes, repeated mini-strokes can, over time, damage the brain. In this case, symptoms develop gradually.

What is mixed dementia?
This term refers to a combination of Alzheimer’s disease and vascular brain damage in the same individual. It happens more frequently in advanced old age.

DIAGNOSIS

When and where is the diagnosis made?
The first step is a consultation with the General Practitioner. After an overall assessment, the GP will decide whether to refer the patient for a specialist consultation (a memory consultation, or to a memory and research centre) or to a professional specialist (neurologist or psychiatrist).

The diagnosis is multidisciplinary. Ideally it will include a neuropsychological assessment, a brain scan, a neurological examination, a comprehensive medical check-up and a psychiatric examination if necessary.

In the case of Alzheimer’s disease and related illnesses, diagnosis may take a long time, mainly because of the gradual nature of the symptoms. The borderline between the benign and the pathological is not always clear at the early stages of the disease.

The average period between the onset of initial problems and a diagnosis is 24 months in France. This delay in diagnosis could be shortened if everyone was vigilant.
What is a neuro-psychological check-up?
This is a series of tests in the form of questions or carrying out simple tasks. The patient’s cognitive impairment is thereby assessed: memory, language, understanding, reasoning, planning, etc.

It is a method which favours the earliest possible detection of symptoms. The tests are adapted to each patient, according to their socio-cultural level in particular, and stage of the disease.

The earlier the test is done, the more accurate it is in highlighting problems which might otherwise go unnoticed.

At the advanced stage of the disease, it is impossible to carry out tests, as the person affected is no longer capable of responding to instructions.

What is brain imaging?
The techniques of brain imaging, scanning and MRI – allow for the examination of brain structures, but not the pathological cells, which are only visible after a post-mortem autopsy* (*An autopsy is not automatic. It may be done to verify a diagnosis or in the context of research).

Until now, these techniques were used to highlight any atrophy affecting certain areas of the brain (the frontal lobe in particular) and to ensure that there were no other pathologies (strokes, tumours).

New techniques are now being used to visualise specific atrophy of certain structures (especially the hippocampus) or abnormalities of blood supply to different areas of the brain.

What does a neurological examination consist of?
The aim is to detect possible neurological disorders in the patient; oculo-motor disorders, trouble with walking, balance problems, parkinsonian syndrome, etc.

At the onset of the disease, the neurological examination is often normal because disorders are not yet apparent and so cannot be detected at this stage. Consequently, an examination alone is not enough to determine that no neurodegenerative disease is present.

What is the “Mini Mental State Examination” (MMSE)?
The “Mini Mental State Examination” (MMSE) is a simple, thirty-item test that rapidly tests overall cognitive function. It is often used by doctors:
- either with the initial aim of referring a patient for a memory consultation
- or with the aim of quickly assessing the progress of the disease, especially in a residential care home for dependent elderly people.

What is the value of a comprehensive medical check-up?
This is essential to ensure that there is no organic illness, no infection (urinary, lung), malnutrition, heart disease or sensory problems (eyesight, hearing).

Any health problems not directly linked to Alzheimer’s disease can contribute to mental confusion and disorientation.

Laboratory tests (blood and urine analyses) are used to screen for pathologies which could lead to cognitive impairment: vitamin or hormone deficiencies, dehydration, infection, intoxication...

In addition, hidden depression in an elderly subject can significantly alter cognitive function. These problems are reversible if suitable treatment is put in place quickly.
Can treatment prevent or delay the onset of the disease?
There is no “miracle” preventive solution but doctors have established a list of recommendations that can delay the risk of developing the disease:

- Preventing cardio-vascular risks (high blood pressure, obesity, diabetes, high cholesterol, smoking)
- A balanced diet
- Physical exercise
- Social and leisure activities

Are there any drugs available to treat the disease?
There are currently no drugs capable of curing the disease or stopping its development.

There are, however, four drugs that can slow the development of memory, language and reasoning problems, and give the initiative back to people affected. The earlier these are prescribed, the more effective they are.

These medicines have a modest success rate and their effects will last for a certain time only although this will vary from patient to patient.

Are there any treatments apart from drugs?
Yes. Non-drug therapies, as part of a “psycho-social” approach, have been the topic of a great many articles and although they have not been subjected to rigorous scientific assessment, their usefulness is widely accepted. It is important that they be considered purely as areas of research.

Different types of help can be recommended for someone depending on the development of their symptoms.

- Creative workshops: painting, sculpture, writing, music...
- Well-being workshops: alternative medicines, tai-chi-chuan...
- Cognitive workshops: memory exercises, reminiscence groups...
- Psychological support.

These types of therapeutic, non-medical help should be provided within a planned package for care and always with trained and qualified professionals: therapists in music and art, speech therapists, psychologists, physiotherapists, etc.
This translation of an original Association France Alzheimer guide “La Maladie d’Alzheimer et les maladies apparentées” was created by the volunteers of Association France Alzheimer Dordogne
www.francealzheimer.org/dordogne
The translation was made possible by a generous donation of the North East Dordogne Women’s Association (NEDWA).
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Writing : France Alzheimer et maladies apparentées
Photo Credit : Cyril Badet, Jean-Louis Courtinat, Olivia Frysowski, Christophe Hargoues, Catherine Thorel, Fotolia/Monkey Business, Pojolaw, Stevepb
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