



## **Rare forms of dementia:**

### Final Report



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# 1. Introduction

The number of people suffering from rare forms of dementia is by definition low and consequently very little has been done to provide information in the same way that it has been for other more common forms such as Alzheimer's disease.

However, when one adds up the number of people in the whole of Europe suffering from a rare form of dementia, there is a sizeable group of people who would benefit from information i.e. on the characteristics of a particular form of dementia, its progression, its treatment possibilities, the problems caregivers face, its prevention...

The aim of this project was to build up a database of information about rare forms of dementia and to make this available to the general public by means of the existing Internet and Intranet systems of Alzheimer Europe. The project would enable quality information on rare forms of dementia to be disseminated throughout Europe.

The description of the diseases was carried out by a group of professionals in order to ensure that the information used was correct and up-to-date. However, it was of primary importance to ensure that the information was written and presented in a style, which was accessible and understandable by the general public.

The working methodology of this project was:

- To elaborate a list of "rare" forms of dementia
- To group these rare diseases in a classification system
- To define the framework of the future database
- To collect quality information
- To analyse the existing information
- To present the information on rare forms of dementia in an easy-to-understand fashion
- To elaborate a glossary to explain specific words
- To build up the Rare Forms of Dementia Database
- To insert this collected information into the database

## 2. Expert group

Alzheimer Europe created an expert group made up of different member organisations of Alzheimer Europe and professionals. This group was established in order to ensure that the information used was correct and up-to-date as well as to ensure that the information database was widely circulated in the different member States of the European Union.

Hereafter you will find the list of persons involved in this expert group.

For full details, please see **Annex 1**.

- Kurt Jellinger (Austria)
- André Delacourte (France)
- Alexander Kurz (Germany)
- Giuliano Binetti (Italy)
- Jos Van der Poel (The Netherlands)
- Clive Evers (United Kingdom)

Unfortunately, Gunhild Waldemar (Rikshospitalet, Copenhagen) was unable to participate in the project. But luckily Giuliano Binetti, a neurologist working for the Alzheimer Unit at Brescia (Italy), agreed to be a part of the expert group.

## 3. Experts' meetings

The first expert group meeting was held in Luxembourg on 17-19 January 2003. During this meeting, the experts had to agree on the establishment of a general structure for the database of information about rare forms of dementia: to elaborate a list of "rare" diseases, to classify these diseases, to elaborate relevant categories of information and to collect quality information.

A second meeting was organised in Luxembourg on 17-18 May 2003. It was mainly dedicated to the analysis of the descriptions of the diseases and to improve the classification system and the descriptive categories (Please see **Annex 2** for the agenda and the minutes of the two meetings).

## 4. Collection of existing information

### 4.1. Lists of documents collected from member organisation of Alzheimer Europe

First we analysed certain publications of our member organisations. Some countries had information relevant to the project, whereas others had none. In general, there was very little written material available on this subject. Hereafter you will find the list of the existing documents.

Country	Association	Brochures	Language
Finland	Alzheimer-keskusliitto	Vaskulaarinen dementia	Finnish
Finland	Alzheimer-keskusliitto	Lewyn kappale-tauti	Finnish
Finland	Alzheimer-keskusliitto	Frontaalinen eli otsalohkodementia	Finnish
Sweden	Demensförbundet	Vad är demen?	Swedish
Sweden	Demensförbundet	Demens-sjukdomen	Swedish
Sweden	Demensförbundet	Rättighets fonden	Swedish
The Netherlands	Alzheimer Nerderland	Lewy body dementie	Dutch
The Netherlands	Alzheimer Nerderland	Vasculaire dementie	Dutch
The Netherlands	Alzheimer Nerderland	Ziekte van Pick	Dutch
United Kingdom	Alzheimer's Disease Society	Creutzfeldt-Jakob Disease	English
Country	Association	Articles	Language
Sweden	Demensförbundet	Doktorn har ordet Frontallobsdemens <i>Article extract from Demens-Forum NR4-2002</i>	Swedish
Sweden	Demensförbundet	Tröst till personer med svar demens <i>Article extract from Demens-Forum NR1-2003</i>	Swedish
Switzerland	Association Alzheimer Suisse	L'évolution d'une paralysie supranucléaire progressive <i>Article extract from Alzheimer Info 29</i>	French, German
Switzerland	Association Alzheimer Suisse	L'évolution d'une aphasie progressive primaire <i>Article extract from Alzheimer Info 30</i>	French, German
Switzerland	Association Alzheimer Suisse	L'évolution d'une démence avec corps de Lewy <i>Article extract from Alzheimer Info 31</i>	French, German
Switzerland	Association Alzheimer Suisse	Maladie de Creutzfeldt-Jakob <i>Article extract from Alzheimer Info 32</i>	French, German

Country	Association	Information sheets	Language
The Netherlands	Alzheimer Nerderland	De ziekte van Creutzfeld-jakob	Dutch
The Netherlands	Alzheimer Nerderland	Lewy body ziekte	Dutch
The Netherlands	Alzheimer Nerderland	Ziekte van Parkinson en dementie	Dutch
The Netherlands	Alzheimer Nerderland	Ziekte van Pick	Dutch
The Netherlands	Alzheimer Nerderland	Vasculaire dementie	Dutch
The Netherlands	Alzheimer Nerderland	Het Korsakov syndroom	Dutch
United kingdom	Alzheimer Scotland - Action on Dementia	Dementia with Lewy bodies	English
United Kingdom	Alzheimer Scotland - Action on Dementia	Vascular dementia	English
United Kingdom	Alzheimer's Disease Society	Dementia with Lewy bodies	English
United Kingdom	Alzheimer's Disease Society	What is Korsakoff's syndrome?	English
United Kingdom	Alzheimer's Disease Society	Rare causes of dementia	English

## 4.2. Information obtained on Internet

We had also established an inventory of existing information on rare forms of dementia available on the Internet and extensive material was obtained from this source. However it is important to stress that the majority of this available information was written in a medical language and not adapted to the general public. (Please see **Annex 3** for a list of material obtained).

## 5. "Rare" forms of dementia

According to the programme of community action on rare diseases:

*"For the purposes of this programme, rare diseases, including those of genetic origin, are life-threatening or chronically debilitating diseases which are of such low prevalence that special combined efforts are needed to address them so as to prevent significant morbidity or perinatal or early mortality or a considerable reduction in an individual's quality of life or socio-economic potential;*

*As a guide, low prevalence can be understood as meaning prevalence which is generally recognised as less than **5 per 10 000** in the Community."*

On the basis of the general epidemiological data available, people who have dementia account for more or less 1% of the global population in the European community <sup>1</sup>.

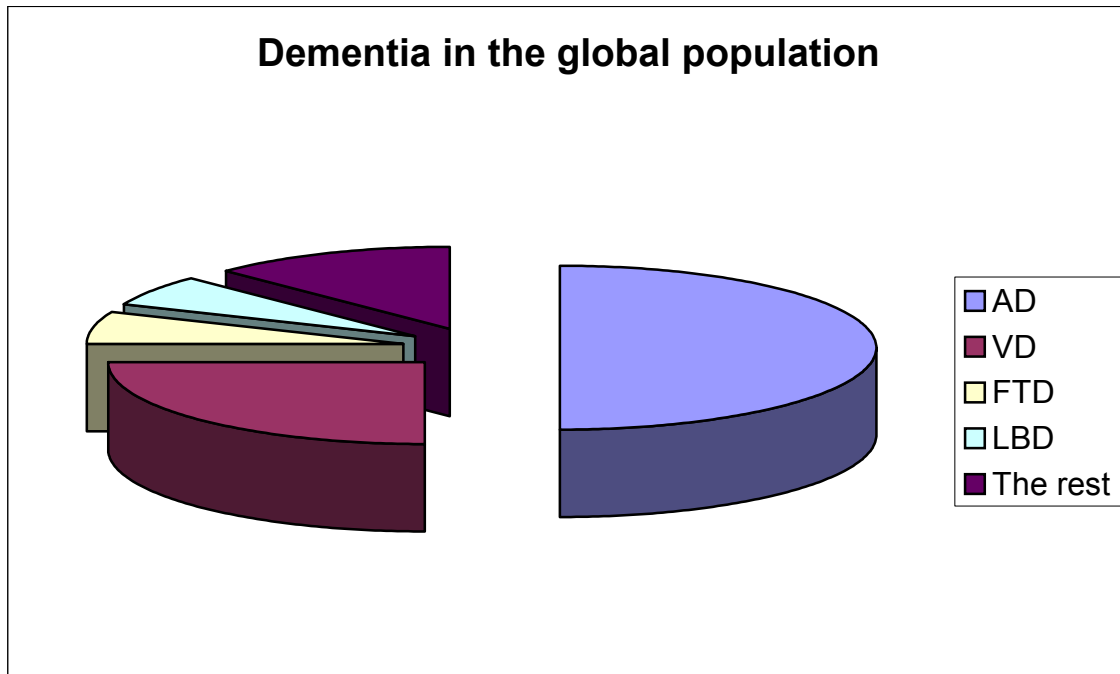
It is generally accepted that the majority of dementias are caused by Alzheimer's disease (approximately 50%) and Vascular Dementias (approximately 25%). For this reason, these "common" dementias were not covered in this project. However the expert group decided to add a section on the rare variants of the common causes of dementias.

The expert group also decided to limit the rare diseases to forms of dementia occurring in adults (the ones appearing in infants and children will be not covered in this project).

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<sup>1</sup> Hofman, A. et al. (1991), The prevalence of dementia in Europe: a collaborative study of 1980-1990 findings, International Journal of Epidemiology, Volume 20, No.3, pages 736-748.

People having ...	Epidemiological data
Dementia in general	100/10.000
Alzheimer’s disease (AD) (+/- 50% of the global population having dementia)	50/10.000
Vascular dementias (VD) (+/- 25% of the global population having dementia)	25/10.000
Fronto-temporal degeneration (FTD) including Pick’s disease, Primary Progressive Aphasia,...	6,25/10.000
Lewy Body Diseases (LBD) including Lewy Body Dementia and dementia in Parkinson’s disease (+/- 6,25% of the global population having dementia)	6,25/10.000





## 6. Classification systems

Working from the epidemiological data agreed upon, the group of experts established a classification system during the first meeting for the rare forms of dementia.

In order to improve the classification system, the expert group decided to add a new sub-category called "Cerebro-vascular diseases". This category will cover rare forms of vascular dementias such as CADASIL, Binswanger disease, Amyloid angiopathy and Subdural hematoma. Other rare forms of dementia were also included in this classification system.

Hereafter you will find the list of the different diseases included in the Classification system.

Name	Category	Person in charge
1. Familial British Dementia	Degenerative	André Delacourte
2. Postencephalitic Parkinsonism	Infectious	Kurt Jellinger
3. Hallervorden-Spatz	Metabolic	Kurt Jellinger
4. Herpes Encephalitis	Infectious	Kurt Jellinger
5. Dementia in hepatic and renal failure	Metabolic	Kurt Jellinger
6. Subdural hematoma	Cerebro-vascular	Kurt Jellinger
7. Dementia in chronic intoxications	Toxic	Kurt Jellinger
8. Drug induced dementia	Toxic	Kurt Jellinger
9. Chronic hypovitaminoses	Metabolic	Kurt Jellinger
10. Down syndrome	Degenerative	Jos Van der Poel
11. GSS and FFI	Infectious	André Delacourte

During the second meeting, other changes were also suggested by the expert group such as:

- To include Familial Autosomal Dominant AD under Degenerative disease as N° 1
- To rename the category entitled "Other rare diseases causing dementia" by "Other rare causes of dementia"
- To rename "Lewy Body Spectrum" by "Lewy Body Disease"
- To rename "Fronto-temporal dementia" by "Fronto-temporal degeneration"

For the full reviewed Classification system, please see **Annex 4**.

## **7. Framework of the database of information**

### **7.1. General Introduction**

In order to improve the understanding of the rare forms of dementia, the expert group decided to elaborate a "General Introduction". The objective of this introduction was mainly to assemble basic information common to all the rare forms of dementia analysed within the framework of our project.

The general introduction is made up of 5 specific sections presenting general information on:

1. Dementia : definition and concept
2. The functioning of the brain
3. The diagnostic procedures
4. Genetic causes of dementia
5. Care and treatment of dementia including the patient perspectives, the caregiver problems, the financial aspects, the legal aspects, the drug treatment and non-drug treatment.

The expert group agreed on the importance that the information should be written and presented in a style that will ensure that it is accessible and understandable by the general public.

## 7.2. Descriptive categories

In order to have comparative information, the experts stressed the importance that all the rare forms of dementia should be described under the same descriptive categories. However, these categories were directly linked to the needs of the target audience.

The target audience consisted of a variety of people such as caregivers, family members, family associations, patients and general practitioners. The needs of information were different from one group to another. According to the differences, the experts decided to elaborate two major sections:

For the caregivers, family members, family associations and patients, the information is supposed:

- To improve the understanding of other forms of dementia than Alzheimer's disease
- To help family members and caregivers to better understand the patient
- To reduce caregivers' burden and family conflicts
- To improve the quality of care

For the general practitioners, the information is supposed:

- To facilitate identification of non-Alzheimer and non-Vascular dementias
- To improve the differential diagnosis
- To ensure the identification of treatable causes
- To avoid ineffective and expensive treatment

On the basis of this analysis, descriptive categories were proposed during the first meeting and reviewed during the second meeting. In order to avoid oversimplifying too much the diseases' information, the experts decided to elaborate a glossary that will explain the more technical words.

Hereafter you will find the **final descriptive categories**.

1. General outline (Ensure that dementia/cognitive impairment are included)
2. Synonyms
3. Symptoms and Course Caregiver problems (only if these are specific)
4. Causes and risk factors
5. Genetics
6. Diagnostic procedures
7. Care and treatment
8. Frequency
9. Ongoing research / Clinical trials
10. Available services
11. References

## 8. Final document

All the information on the rare forms of dementia and the general introduction are now available on Alzheimer Europe Internet site under the section "Other forms of dementia". Here is the URL address to consult this available information:

<http://www.alzheimer-europe.org/?lm1=D76EBF7F6AEA>

The advantages of the Internet tool are that detailed information can be accessible world wide and constantly updated.

The Alzheimer Europe Internet site gives also the opportunity to the users to print an article on specific diseases without banners or to email it to a friend.

The glossary is also consultable on Alzheimer Europe Internet site by clicking on "View Glossary" button and let the opportunity to the user to find a specific word. This glossary is also directly linked to the articles presenting the diseases' information and when a word is defined in the glossary, it will be highlighted in blue. When the user moves his/her cursor over a word, the definition will appear automatically in a pop-up window.

Please see the final document, including the General Introduction, the description of all the diseases and the glossary herewith enclosed.

**Annex 1** : List of participants

**Annex 2** : Agenda and minutes of the 2 Experts' meetings

**Annex 3** : Information obtained on Internet

**Annex 4** : Classification system

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