Epidemiology of the different Clinical Presentations of Early Onset Dementia

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BACKGROUND

Patients with Early Onset Dementia (EOD), defined as dementia with symptoms onset before the age of 65, frequently present with atypical clinical syndromes.

However, the epidemiology of the different presentations of EOD, including the clinical variants of Alzheimer's dementia (AD) and frontotemporal dementia (FTD), has never been investigated altogether in the same population-based study, probably because their characterization is relatively recent, and subsequent to the advent of AD biomarkers.

Epidemiologic data on all-causes of EOD are also scarce, inconsistent, and mostly based on low-level clinical assessment.

We investigated EOD epidemiology by identifying all EOD patients seen in the extended network of dementia services of the Modena province, Northern Italy (around 700,000 inhabitants):
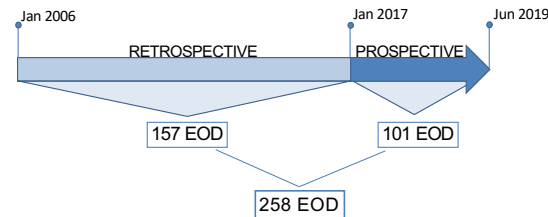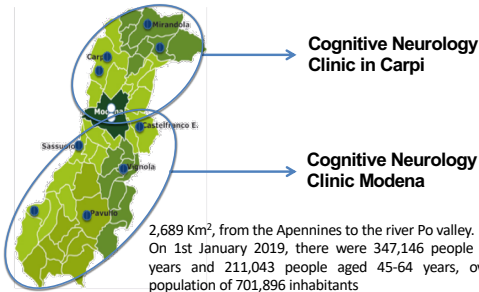- All-causes EOD prevalence and incidence
- EOD clinical phenotypes prevalence and incidence

METHODS

PATIENTS RECRUITMENT: all newly-diagnosed EOD patients in the province of Modena in the period January 2006 – June 2019

NETWORK OF DEMENTIA SERVICES:
- 2 hospital-based outpatient Cognitive Neurology clinics
- 8 outpatient geriatric Memory clinics (Centro per i disturbi cognitivi e le demenze, CDCD).

Cognitive Neurology Clinic in Carpi

Cognitive Neurology Clinic Modena

2,689 Km², from the Apennines to the river Po valley.
On 1st January 2019, there were 347,146 people aged 30-64 years and 211,043 people aged 45-64 years, over a total population of 701,896 inhabitants

RESULTS

Jan 2006 — RETROSPECTIVE — Jan 2017 — PROSPECTIVE — Jun 2019

157 EOD | 101 EOD

258 EOD

Incidence

From 1st January 2016 to 30th June 2019: 160 incident cases with a median age of onset of 60 years

All-causes EOD crude and age- and sex-adjusted incidence: 6.5/100000 inhabitants/year

Corresponding to 46 new cases per year

Stratifying the analyses for age group:
- adjusted incidence 13.2 in 30-64 age group
- adjusted incidence 22.1 in 45-64 age group

Prevalence

At 30th June 2019 all-causes EOD crude and age and sex-adjusted prevalence: 36.4/100000 inhabitants

Stratifying the analyses for age group:
- adjusted prevalence EOD 74.3/100000 in 30-64 age group
- adjusted prevalence of EOD was 119.8/100000 in 45-64 age group

Diagnoses

Clinical diagnosis	N	%	M/F
AD	113	43.8	36/77
FTD spectrum	78	30.2	44/34
Vascular dementia	24	9.3	16/8
Lewy bodies dementia	9	3.5	5/4
Leukoencephalopathy	7	2.7	5/2
Parkinson's disease dementia	7	2.7	6/1
Alcoholic dementia	4	1.5	3/1
Cerebral amyloid angiopathy	3	1.2	2/1
Dementia in Huntington disease	3	1.2	2/1
Not specified	10	3.9	4/6

CLINICAL DIAGNOSIS OF PREVALENT CASES
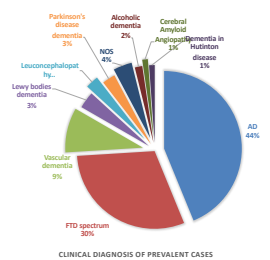
Clinical Subtypes

Clinical subtypes	Crude prevalence (per 100,000)	Crude prevalence per 100,000 persons at risk (aged 30-64)	Crude incidence (per 100,000)	Crude incidence per 100,000 persons at risk (aged 30-64)
AD	**16.0** (113)	**32.6**	**2.5** (61)	**5.0**
amnestic	**10.6** (75)	**21.6**	**1.7** (43)	**3.5**
Posterior cortial atrophy (PCA)	**1.1** (8)	**2.3**	**0.08** (2)	**0.16**
Logopenic variant (lvPPA)	**2.8** (20)	**5.8**	**0.4** (10)	**0.8**
Behavioural/disexecutive	**0.7** (5)	**1.4**		
FTD SPECTRUM	**11.0** (78)	**22.5**	**2.1** (52)	**4.3**
Behavioural variant (bvFTD)	**6.9** (49)	**14.1**	**1.3** (32)	**2.6**
Semantic variant (svPPA)	**1.4** (10)	**2.9**	**0.3** (8)	**0.66**
Non fluent primay progressive aphasia (nfvPPA)	**0.3** (2)	**0.6**	**0.08** (2)	**0.16**
FTD-MND (all SLA-FTDs)	**0.8** (6: 4 bvFTD, 2nfvPPA)	**1.7**	**0.16** (4: 3bvFTD, 1nfvPPA)	**0.32**
Corticobasal syndrome (CBS)	**0.7** (5)	**1.4**	**0.08** (2)	**0.16**
Progressive sopranuclear palsy (PSP)	**0.6** (4)	**1.2**	**0.12** (3)	**0.25**

CONCLUSIONS

- The most frequent cause of EOD was AD
- The most frequent EOD phenotype was the amnestic variant of AD, followed by the behavioural variant of FTD, and by the logopenic variant of AD
- Considering all the AD cases, the non-amnestic variants (i.e. lvPPA, PCA and bdvAD) were 34% of all AD cases, with lvPPA being the most frequent.
- Knowing the relative frequency of presentations will directly benefit the reasoning of clinicians faced with young patients with cognitive symptoms
- The result of this population study may contribute to a better understanding of epidemiology and clinical management of EOD, and its different clinical variants, thus helping to optimize cost-effective dementia care organization, and ultimately to improve patients' and caregivers' quality of life