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Effect of diagnostic criteria on prevalence of frontotemporal dementia in the elderly.

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Abstract

Background: Frontotemporal dementia (FTD) is believed to be rare in the elderly, and the influence of different criteria on the prevalence of FTD is unclear.

Methods: Population-based samples of 70 to 95-year-olds (n =2462) in Gothenburg, Sweden underwent neuropsychiatric examinations. Behavioral variant FTD (bvFTD) was diagnosed according to the International bvFTD Criteria Consortium (FTDC), the Frontotemporal Lobe Degeneration Consensus Criteria and the Lund-Manchester Research Criteria. A subset (n=1074) underwent CT of the brain.

Results: The prevalence of bvFTD varied between 0.2-0.5% at age 70-79, between 2.5-3.6% at age 80-89, and between 1.7-2.2% at age 90-95. The agreement between different criteria was low to moderate (kappa=0.20-0.42). Among those with bvFTD according to FTDC, 93.3% had frontal and/or temporal lobar atrophy on CT, compared to 12.6% of those without bvFTD (p<0.001).

Conclusions: The prevalence of bvFTD was higher than expected in this population. To a large extent, different criteria captured different individuals.

1. Introduction

The diagnosis of frontotemporal dementia (FTD) continues to be challenging for clinicians and researchers.¹ Among the three subtypes of FTD, the most common is the behavioral variant FTD (bvFTD), while other subtypes (semantic dementia and primary progressive aphasia) are rare.^{1,2} The core clinical manifestations of bvFTD are progressive deterioration of personal and social conduct, and emotional and motivational blunting. Memory loss, apraxia, agnosia and impaired spatial orientation most often occur late in the disease. Cases of bvFTD may thus not initially fulfill current criteria for dementia, where memory impairment is mandatory.^{3,4} Frontal lobe symptoms may also be caused by trauma and disorders such as Alzheimer's disease, vascular dementia, frontal lobe tumors and alcohol-associated dementia.

There are currently three clinical criteria sets for the diagnosis of bvFTD. The first criteria, Lund-Manchester Research Criteria (LMRC), were published in 1994,⁵ followed in 1998 by the Consensus Criteria for Frontotemporal Lobar Degeneration (FTLD-CC).⁶ In 2011, the International Behavioral Variant FTD Criteria Consortium (FTDC) proposed revised criteria,¹ as the 1998 criteria were considered to be too rigid for clinical and research purposes.⁷ These three sets of diagnostic criteria include different combinations of impairments in social and emotional abilities.

Few population studies have examined the frequency of FTD among the elderly. Most epidemiological studies have been performed in the age group 45-65 years,⁸ in which prevalence estimates range from 2.0-15.4 per 100,000.^{9,10,11,12,13} In individuals above age 65 years, most studies report that the prevalence is below one percent (using LMRC or FTLD-CC criteria).^{14,15,16,17} An exception is a study from Italy reporting a prevalence of 5.2% in individuals aged above 70 years.¹⁸ However, this study was conducted in an isolated

population with a high frequency of hereditary FTD. Furthermore, the studies among the elderly only included cases of FTD who also fulfilled criteria for global dementia, in which memory problems are mandatory. Thus, individuals with FTD who do not fulfill criteria for global dementia may remain undetected.^{19,20} Furthermore, key informant interviews (with close relatives and caregivers) were used in only two of the studies among the elderly.^{14,17} Key informant interviews are crucial to obtain retrospective information about early symptoms and course of symptoms, as these are necessary to differentiate bvFTD from other dementia disorders. Using a combination of clinical examinations and key informant interviews, a prevalence of 3% for bvFTD according to the LMRC was reported in a representative population of 85-year-olds from Gothenburg, Sweden.²¹ Thus, while it has been suggested that FTD may be more common than previously supposed,^{22,23,24} few studies have examined the prevalence of FTD in a wider range of ages among the elderly. Neither has the utility of different criteria been examined in elderly populations.

Our aim was to examine the prevalence of bvFTD in population samples of 70-95-year-olds from Gothenburg, Sweden, using three sets of criteria (the FTDC, the FTLD-CC and the LMRC), and to determine the agreement between these criteria.^{1,5,6} A further aim was to study the correlation between bvFTD and the occurrence of frontal and/or temporal lobe atrophy on computerized tomography (CT) of the brain.

2. Subjects and methods

2.1. Subjects

Between 1986 and 2001, studies on representative elderly populations in Gothenburg, Sweden were conducted using identical examinations (including neuropsychiatric examinations and key informant interviews) at each occasion.²⁵ All samples were systematically obtained from the Swedish population register based on birth dates, and included people living in private households and in residential care. To examine the age-specific prevalence of bvFTD, we merged data from these studies.

The H70-study: In 2000-01, an effective sample of 827 70-year-olds was selected and a total of 579 individuals (350 women and 229 men) agreed to participate (response rate 70%).²⁶ There were no differences between participants and non-participants regarding sex, marital status or previous outpatient or inpatient psychiatric care. Non-participants had higher five-year mortality rate than participants both among women (9.0% vs. 2.3% $p<0.001$) and among men (23.7% vs. 7.5%, $p<0.001$), as described previously.²⁶

The H85-study. In 1986-7, an effective sample of 783 85-year-olds was selected and a total of 494 individuals (351 women and 143 men) agreed to participate (response rate 63%).²⁷ There were no differences between participants and non-participants regarding sex, marital status, registration as psychiatric outpatients or inpatients, three-year mortality rate and institutionalization. Identical studies in this sample were conducted at ages 88 ($n=260$), 90 ($n=200$) and 92 years ($n=190$).^{27,28}

The 95+ study. In 1996-98, an effective sample of 529 95-year-olds was selected and a total of 338 individuals (263 women and 75 men) agreed to participate (response rate 64%). There

were no significant differences between participants and non-participants regarding marital status and three-year mortality rate.²⁹

The Prospective Population Study of Women. In 1992-93, an effective sample of 837 women (aged 70, 74, 78 and 84) was selected and a total of 559 women (response rate 67%) agreed to take part (255 aged 70, 215 aged 74, 70 aged 78 and 19 aged 84).^{30,31,32,33} In 2000-2001, 629 of the women were alive, and 439 (response rate 70%) agreed to participate in neuropsychiatric examinations (216 aged 78, 171 aged 82, 44 aged 86 and 8 aged 92).

The data from these studies were merged, and 630 individuals without key informant interviews were excluded, leaving 2462 (79.6%) for study (503 men, 1959 women). The merged sample was stratified by ages 70-79, 80-89 and 90-95 (table 1).

The Ethics Committee for Medical Research at Gothenburg University approved all studies. Informed consent was obtained from the participants, their nearest relatives, or both.

2.2. Methods

Identical neuropsychiatric examinations and key informant interviews were used for all participants included in this study. The neuropsychiatric examinations were semi-structured and performed by trained neuropsychiatrists, except in 2000-2001 when they were performed by experienced psychiatric nurses. The examinations included ratings of symptoms and signs common in dementia and a cognitive test battery.²⁷ Psychiatric symptoms and signs were rated with the Comprehensive Psychopathological Rating Scale.³⁴ Frontal lobe symptoms assessed included disinhibition, aggressiveness, hyperorality, hyperphagia, hypersexuality,

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perseverative or stereotypic behavior, utilization behavior, apathy, emotional bluntness and loss of empathy. Tests of cognitive function included assessments of recent and remote memory, orientation for time, place, person and situation, apraxia, constructional apraxia, ideational apraxia, ability to understand proverbs, ability to follow commands, finger agnosia, judgment, and language. The Mini Mental State Examination³⁵ and a global rating of mental health were also performed.

The semi-structured telephone-interviews with key informants included questions about cognitive, emotional and behavioral symptoms, e.g. global changes in personality, memory, orientation, difficulties in finding way in familiar surroundings, intellectual ability, language, speech, motivation, disinhibition, emotional bluntness, suspiciousness and paranoid ideas, depression, lachrymosity, anxiety and worries, irritability, aggressive behavior, performances in activities of daily living, and insight. Questions were asked about age at onset and course of symptoms.²⁷ The retrospective information from key informants was needed to elucidate early symptoms and course of symptom development. The data were collected blindly to any diagnostic aspects.

All interviewers were trained and supervised by the last author (IS), who also performed the examinations of 85-year-olds in 1986-87.

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2.3. Computerized tomography (CT) of the brain

A systematic subsample of 1900 individuals was invited to undergo CT-scanning of the head, and 1074 accepted (244 men, 830 women). Of these, 161 had global dementia as diagnosed by DSM-III-R (41 men, 120 women), and 913 were without dementia (203 men, 710 women).

All CT-scans were performed without contrast enhancement and with 10 mm continuous slices. The CT-scans were evaluated either by radiologists or a neurologist experienced in rating CT-scans. The evaluations were done blindly to the results of the neuropsychiatric examination. Location of cortical atrophy was categorized as frontal, temporal, parietal or occipital, according to the anatomical subdivision.³⁶ A scale with three grades (absent vs. mild vs. moderate or severe) was used to estimate cortical atrophy according to the extent of sulcal widening.³⁷ Inter-rater agreement for the assessment of atrophy was “fair” for frontal lobe atrophy (kappa = 0.34) and “moderate” for temporal lobe atrophy (kappa = 0.43).³⁷ The intra-rater kappa values for the assessment of atrophy were “moderate” for frontal lobe atrophy (kappa = 0.53) and “good” for temporal lobe atrophy (kappa = 0.61).³⁸

2.4. Diagnostic procedures

The FTDC criteria for bvFTD define five symptom clusters: disinhibition, perseveration, apathy, lack of empathy and hyperorality. Symptoms from at least three clusters need to be present for a diagnosis.¹ The algorithm based on the FTDC is described in figure 1.

The 1998 consensus criteria (FTLD-CC) for bvFTD define four frontal lobe symptom clusters: impaired social conduct, impaired personal conduct, emotional blunting and loss of

insight. For a diagnosis of bvFTD, it is mandatory to have symptoms from all symptom clusters.⁶ The algorithm based on the FTLD-CC is described in figure 2.

The LMRC define three clusters of frontal lobe symptoms (behavioral, affective and language).⁵ Language disturbance is not included as a symptom in FTDC and FTLD-CC criteria for bvFTD. To make criteria comparable with FTDC and FTLD-CC criteria, we therefore only used the first two clusters from LMRC to define a 'LMRC bvFTD' (figure 3).

The first step in the diagnostic process was to select individuals fulfilling symptom criteria, as described above.^{1,5,6} Second, these individuals were evaluated regarding initial symptoms, course, and additional information needed to diagnose or exclude bvFTD (figures 1, 2 and 3). Frontal lobe symptoms had to precede severe amnesia or loss of spatial skills for a diagnosis of bvFTD. Likewise, the course of the symptom clusters had to be compatible with bvFTD with insidious onset and a progressive, non-episodic course. The final diagnosis was reached by consensus between two of the authors (TBG, MSj.). The different bvFTD diagnoses were set blindly of any other dementia diagnoses, and blindly of each other.

Global dementia was diagnosed according to the Diagnostic and Statistical Manual of Mental Disorders, Third edition, revised (DSM-III-R) criteria,³⁹ as described previously.²⁷

2.5. Statistical methods

Pearson's chi-square (χ^2) or, if necessary, Fisher's Exact Test, were used to test differences in proportions. All p-values were two-tailed and p-values <0.05 were considered statistically significant. Cohen's un-weighted kappa was used to assess agreement between different criteria.

3. Results

The prevalence of bvFTD varied between 0.2-0.5% at age 70-79, between 2.5-3.6% at age 80-89, and between 1.7-2.2% at age 90-95 using the different criteria (table 2).

Agreement between criteria was low to moderate with kappa values ranging from 0.20-0.42 (table 3 and figure 4). Only 7 out of 88 bvFTD cases diagnosed with at least one set of criteria were captured by every set of criteria, and 65 were diagnosed according to only one criteria set.

Among bvFTD cases, 79% of those diagnosed by FTDC, 92% of those diagnosed by FTLDC, and 53% of those diagnosed by LMRC had dementia according to DSM-III-R ($p < 0.05$ FTDC vs. LMRC; $p > 0.05$ FTDC vs. FTLDC; $p < 0.05$, FTLDC vs. LMRC).

3.1. Computerized tomography (CT) of the brain

CT-scan was performed in 1074 individuals. Among these, 1.4% ($n=15$) had bvFTD according to FTDC, 1.0% ($n=11$) according to FTLDC and 0.7% ($n=8$) according to LMRC.

Furthermore, 89.4% ($n=960$) of the participants in the CT examination did not have frontal lobe symptoms as defined by any of the three criteria sets (no frontal lobe symptoms; non-FLS). In this group the prevalence of moderate-severe frontal atrophy was 8.9% ($n=85$) and out of these 11.8% ($n=10$) had global dementia as diagnosed by DSM-III-R. Furthermore, in the non-FLS group, the prevalence of moderate-severe temporal atrophy was 8.1% ($n=78$) and 19.2% ($n=15$) of these had global dementia. In the non-FLS group the prevalence of moderate-severe frontal and/or temporal atrophy was 4.4% ($n=42$) with 42.8% ($n=18$) having global dementia.

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The association between frontal and/or temporal lobe atrophy and the bvFTD-diagnoses is shown in table 4. Among 15 cases with bvFTD, 14 (93.3%) had moderate-severe frontal and/or temporal atrophy compared to 12.6% in the non-FLS group ($p<0.001$).

Among 11 persons with bvFTD according to FTLD-CC, 7 (63.6%) had moderate-severe frontal and/or temporal atrophy ($p<0.001$ compared to non-FLS).

All 8 persons with bvFTD according to LMRC had moderate-severe frontal and/or temporal atrophy ($p<0.001$ compared to non-FLS).

Dementia severity as measured by MMSE was not significantly associated with bvFTD as diagnosed by any diagnostic criteria (FTDC: $p=0.791$; FTLD-CC: $p=0.066$; LMRC: $p=0.353$).

Furthermore, dementia severity as measured by MMSE was not significantly associated with frontal and/or temporal lobe atrophy (FTDC: $p=0.301$; FTLD-CC: $p=0.101$; LMRC: $p=0.333$).

4. Discussion

We examined the prevalence of possible bvFTD in an elderly general population using three different FTD criteria. Irrespective of criteria used, bvFTD was more common among the elderly than previously assumed. Furthermore, we found a higher prevalence at age 81-95 than at age 70-79 years. Despite a similar prevalence, the agreement between the three criteria sets was only low to moderate. We also found that a large proportion of those diagnosed according to the FTDC and LMRC (93-100%) had moderate-severe frontal and/or temporal lobe atrophy on CT, which could be compared to 13% in those without frontal lobe symptoms.

It is believed that FTD occurs mainly among individuals aged 45–65 years.^{23,24} and most prevalence studies have been performed in this age group.⁸ In populations above age 65 years, most studies have reported lower rates (0-0.6%)^{14,15,16,17} than our study. One explanation may be that we applied bvFTD criteria directly to the population without prior screening for global dementia. Another reason may be the use of comprehensive key informant interviews to gather information on frontal lobe symptoms and the early course of the disease. Prevalence estimates for the elderly based on register data are lower (only 4-54 per 100 000), probably reflecting that many cases of FTD are not detected by the health care system, or that they receive other diagnoses than FTD.^{19,20} The diagnosis of bvFTD is probably even more underestimated in the very old, as this diagnosis is seldom considered in this age group.^{22,23} Despite a similar prevalence using different criteria, the agreement between criteria was only low to moderate. Among those with bvFTD according to the FTDC, only 27% had a diagnosis according to FTLD-CC, and 35% according to LMRC (with kappa values 0.30-0.42). The LMRC and FTLD-CC had an overlap of only 19-21%, with a kappa of 0.20. Thus, these criteria captured to a large extent different individuals. The FTLD-CC diagnosed somewhat fewer cases than FTDC, in line with suggestions that these criteria are more rigid.⁷ Furthermore, almost all cases diagnosed with FTLD-CC, a large majority of those with FTDC, but only less than half of those diagnosed with LMRC fulfilled criteria for global dementia. One explanation may be that the LMRC is more weighted towards externalized symptoms, such as loss of inhibition and aggressive behaviors, while the FTLD-CC is more weighted towards negative symptoms, such as apathy and loss of initiative, and the FTLD-CC may thus miss a large proportion of cases with mainly loss of inhibition.⁷ The FTDC seems to be somewhere in-between the other criteria in this regard. Our finding is remarkably similar to reports describing low concordance between different criteria for global dementia.⁴⁰

Few studies have compared the clinical criteria of bvFTD with neuroimaging or neuropathology. One study reported that among 137 cases with frontal lobe degeneration at neuropathological examination, 85% fulfilled FTDC criteria for possible bvFTD and 53% fulfilled FTLN-CC criteria.¹ No previous studies have directly compared FTD criteria with MRI or CT in the setting of a population study. However, reports from memory clinics show that the proportion of patients with clinically diagnosed bvFTD who have frontal and/or temporal atrophy on neuroimaging ranges from 50 to 95%.^{1,41,42,43,44} In our study, a large proportion of those diagnosed with the FTDC and LMRC (93-100%), and a lower proportion of those diagnosed with FTLN-CC (63%), had moderate-severe frontal and/or temporal lobe atrophy on CT. Only 13% of those without frontal lobe symptoms had frontal and/or temporal lobe atrophy. This is analogous to several other brain disorders in the elderly. For example, a large proportion of the elderly population fulfils neuropathological criteria for Alzheimer's disease but lacks clinical symptoms.⁴⁵

Among those with moderate-severe frontal lobe atrophy on CT, 85 individuals did not have frontal lobe symptoms as defined by any of the three criteria sets. Only 12% of those had other dementias. One explanation for this result may be that cortical thinning of the frontal lobes is also found in normal aging,⁴⁶ and in these cases may not lead to detectable frontal lobe symptoms. In addition, we cannot exclude the possibility that prior head trauma may partially explain the presence of frontal and/or temporal lobe atrophy in non-demented individuals.⁴⁷ Furthermore, it has to be emphasized that neuroimaging is a supportive, but not mandatory diagnostic feature of all FTD criteria. Thus, these criteria allow a diagnosis of possible bvFTD in the absence of neuroimaging. However, the high correlation between frontal and/or temporal lobe atrophy and bvFTD according to FTDC and LMRC in our population study is similar to that reported from clinical studies and lends support for the validity of our diagnoses.

Among the strengths of the study is the population-based sample, the use of key informant interviews, and that all individuals were examined with a wide range of psychiatric and neurological variables, including those described in the new FTDC criteria, the FTLD-CC and the LMRC. We were therefore able to design symptom algorithms, even though some data were collected before the criteria were published. A further advantage is that we were able to examine the different criteria in relation to frontal and/or temporal lobe atrophy on CT.

There were also limitations and methodological issues. First, response rates were around 63-70%. Although responders and non-responders were similar regarding a number of background factors, it is possible that they differed regarding frontal lobe symptoms. Second, the data were collected over a long period of time using several different population studies. However, the examination methods were identical over the years, all interviewers and examiners were trained and supervised by the same person (IS), and identical methods for case finding were used. Furthermore, evaluation of data and diagnosis of bvFTD were done by the same neuropsychiatrists (TBG, MSj.) irrespective of year of examination. We cannot, however, exclude the possibility that secular trends in the frequency of FTD might have influenced the results. Third, part of the diagnostic criteria as used in this study required retrospective information from key informants. This was necessary to elucidate early symptoms and course of symptom development. Although information from key informants may be uncertain, it is also used in clinical studies. Fourth, our findings are not validated by neuropathology.²⁴ Even with a high correlation with neuroimaging, it is impossible to exclude that some of the persons with bvFTD might have had atypical Alzheimer's disease (AD), argyrophilic grain disease or some other neurodegenerative or vascular disease. This distinction may be especially difficult to make at very high ages, where AD is common. The prevalence of frontal-predominant AD has been difficult to establish.⁴⁸ Previous studies report

that frontal-predominant AD pathology is found in up to 10% of clinical FTD cases.^{49,50} Fifth, FTD among the elderly may occasionally present as an amnesic state and neuroimaging in these cases often shows focal hippocampal sclerosis.²⁴ The LMRC and FTLDD-CC do not permit a diagnosis of FTD in these cases. The FTDC permits a diagnosis of bvFTD in individuals who present with an amnesic state if psychometric testing is compatible with FTD, i.e. if the psychometric tests show executive deficits with relative sparing of episodic memory and visuospatial functions.¹ This criterion could thus not be applied in our cross-sectional study which uses retrospective information to elucidate the early course of the disorder. Some FTD cases according to FTDC may thus have been missed, leading to underestimation of the prevalence of bvFTD. Furthermore, if this criterion had been applied, the correlation between FTDC and the other criteria would have been even lower. Unfortunately, the neuroimaging technique (CT) employed in this study did not allow us to detect hippocampal sclerosis. Finally, we had a larger refusal rate for CT in those with bvFTD than in the rest of the population, resulting in few individuals in that substudy. The results of neuroimaging should therefore be taken with caution.

In summary, we found a higher prevalence of bvFTD than previously reported. The correlation between the different criteria for bvFTD was low, suggesting that further development of research criteria is required. The findings arouse concerns about the validity of comparisons among studies that use different criteria to diagnose bvFTD. Both the FTDC and the LMRC have a high correlation with frontal and/or temporal lobe atrophy, but only a moderate agreement with each other, suggesting that both FTDC and LMRC could be underestimating the prevalence of bvFTD. These results suggest that any modified FTD criteria should allow for a more flexible combination of frontal lobe symptoms than is possible in the current FTD criteria, as it is important to capture all presentations of FTD.

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In conclusion, our findings point to the need for increased awareness of bvFTD in the elderly, and the need for validation of the criteria used to diagnose bvFTD.

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Table 1. Demographic characteristics of sample.

Age	Men % (n)	Women % (n)	All % (n)	p-value for difference between men and women
70-79	16.4 (175)	83.6 (893)	100 (1068)	<0.001
80-89	22.6 (195)	77.4 (666)	100 (861)	<0.001
90-95	25.0 (133)	75.0 (400)	100 (533)	<0.001
Marital status				
<i>Never married</i>	4.4 (22)	9.7 (190)	8.6 (212)	<0.001
<i>Married</i>	55.7 (280)	16.4 (322)	24.5 (602)	<0.001
<i>Divorced</i>	6.4 (32)	15.1 (296)	13.3 (328)	<0.001
<i>Widowed</i>	23.8 (120)	48.4 (947)	43.3 (1067)	<0.001
<i>N/A</i>	9.7 (49)	10.4 (204)	10.3 (253)	0.658
Proportion with higher education	23.6 (119)	13.2 (258)	15.3 (377)	<0.001
Proportion with dementia* at ages:				
70-79	2.3 (5)	5.0 (45)	4.7 (50)	0.211
80-89	27.7 (54)	34.8 (232)	33.2 (286)	0.063
90-95	37.6 (50)	54.0 (216)	49.9 (266)	0.001

* According to DSM-III-R.

Table 2. The prevalence of the behavioral variant of frontotemporal dementia (bvFTD) between age 70 and 95 years using three sets of criteria.

	FTDC	FTLD-CC	LMRC
Age	%(n)	% (n)	% (n)
70-79 (n=1068)	0.5 (5)	0.3 (3)	0.2 (2)
95% CI	(0.2-1.9)	(0.1-0.8)	(0.1-0.7)
80-89 (n=861)	3.6 (31)	2.5 (22)	2.7 (23)
95% CI	(2.5-5.1)	(1.7-3.8)	(1.8-4.0)
90-95 (n=533)	2.2 (12)	1.7 (9)	2.1 (11)
95% CI	(1.3-3.9)	(0.9-3.2)	(1.1-3.6)

FTDC: Criteria of the International Behavioural Variant FTD Criteria Consortium.

FTLD-CC: Frontotemporal Lobar Degeneration Consensus Criteria.

LMRC: Lund-Manchester Research Criteria.

Table 3. Agreement between LMRC, FTLD-CC and FTDC criteria for behavioral variant frontotemporal dementia (bvFTD).

		FTDC	FTLD-CC	LMRC
		n=48	n=34	n=36
		n (%)	n (%)	n (%)
FTDC	n=48	X	13 (27.1)	17 (35.4)
	Kappa		0.30	0.42
FTLD-CC	n=34	13 (38.2)	X	7 (20.6)
	Kappa	0.30		0.20
LMRC	n=36	17 (47.2)	7 (19.4)	X
	Kappa	0.42	0.20	

FTDC: Criteria of the International Behavioural Variant FTD Criteria Consortium.

FTLD-CC: Frontotemporal Lobar Degeneration Consensus Criteria.

LMRC: Lund-Manchester Research Criteria.

Table 4. Frontal and/or temporal lobe atrophy on CT scan of the brain in relation to a diagnosis of behavior variant frontotemporal dementia (bvFTD) according to different FTD criteria.

	Frontal and temporal lobe atrophy[†] on CT scan of the brain			
	Frontal lobe atrophy	Temporal lobe atrophy	Both frontal and temporal lobe atrophy	Frontal and/or temporal lobe atrophy
bvFTD criteria	% (n)	% (n)	% (n)	% (n)
<i>FTDC</i> (n=15)	80.0 (12)*	73.3 (11)*	60.0 (9)*	93.3 (14)*
<i>FTLD-CC</i> (n=11)	45.4 (5)*	54.5 (6)*	36.4 (4)**	63.6 (7)*
<i>LMRC</i> (n=8)	75.0 (6)*	75.0 (6)*	50.0 (4)*	100 (8)*
Non-FLS (n=960)	8.9 (85)	8.1 (78)	4.4 (42)	12.6 (121)

[†] “Atrophy” refers to moderate or severe lobar atrophy.

FTDC: Criteria of the International Behavioral Variant FTD Criteria Consortium.

FTLD-CC: Frontotemporal Lobar Degeneration Consensus Criteria.

LMRC: Lund-Manchester Research Criteria.

Non-FLS: Participants in the CT examination who did not have frontal lobe symptoms as defined by any of the three criteria sets (FTDC, FTLD-CC or LMRC).

*p<0.001 for difference between FTD diagnosis and non-FLS.

**p=0.001 for difference between FTD diagnosis and non-FLS.

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7. Keywords: Frontotemporal dementia; Aged; Prevalence; Tomography, X-Ray Computed.

Figure 1. Algorithm for diagnosis of bvFTD from International consensus criteria for behavioral variant FTD (FTDC).

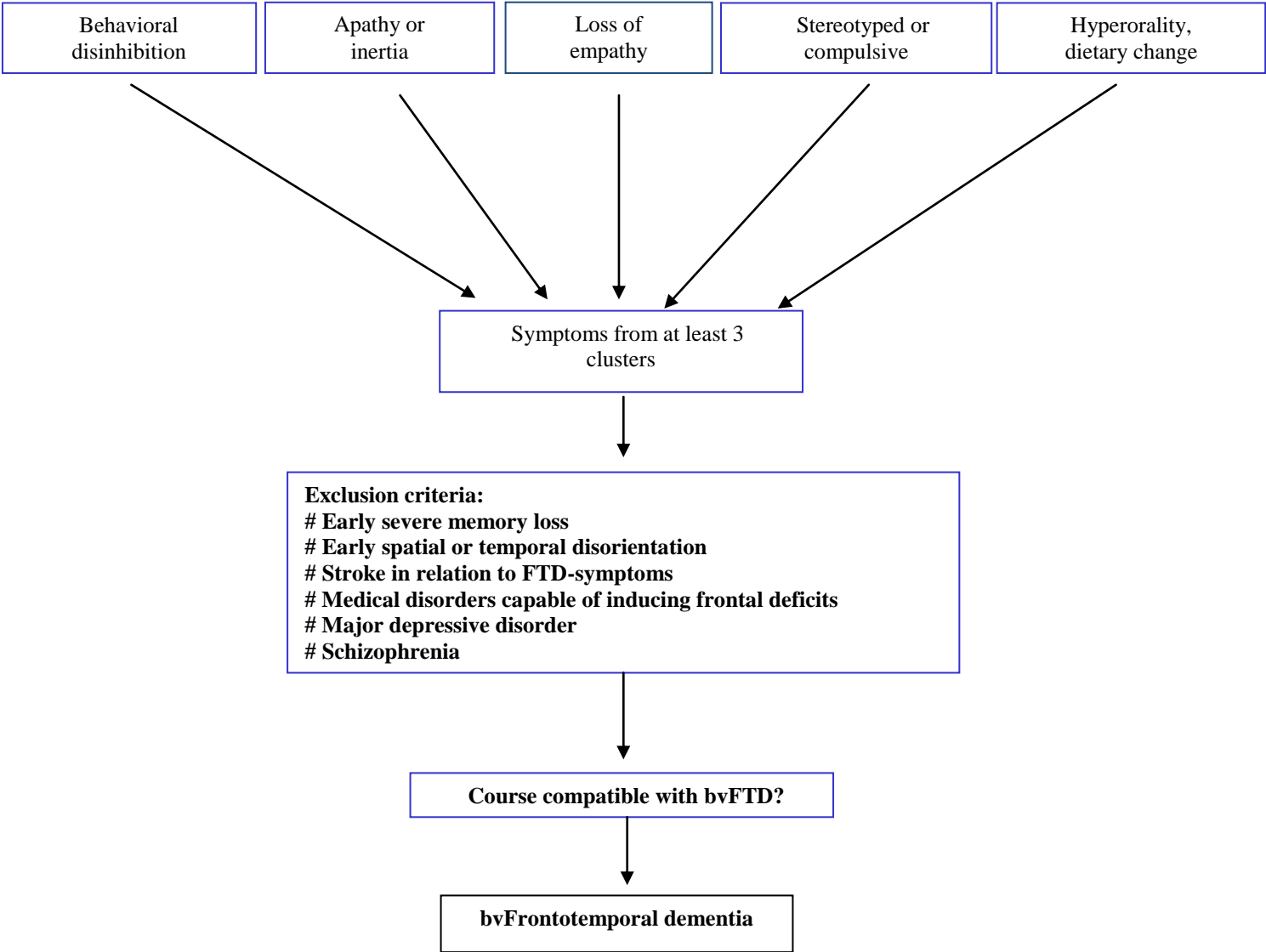


Figure. 2. Algorithm for diagnosis of bvFTD from FTLN consensus criteria (FTLN-CC).

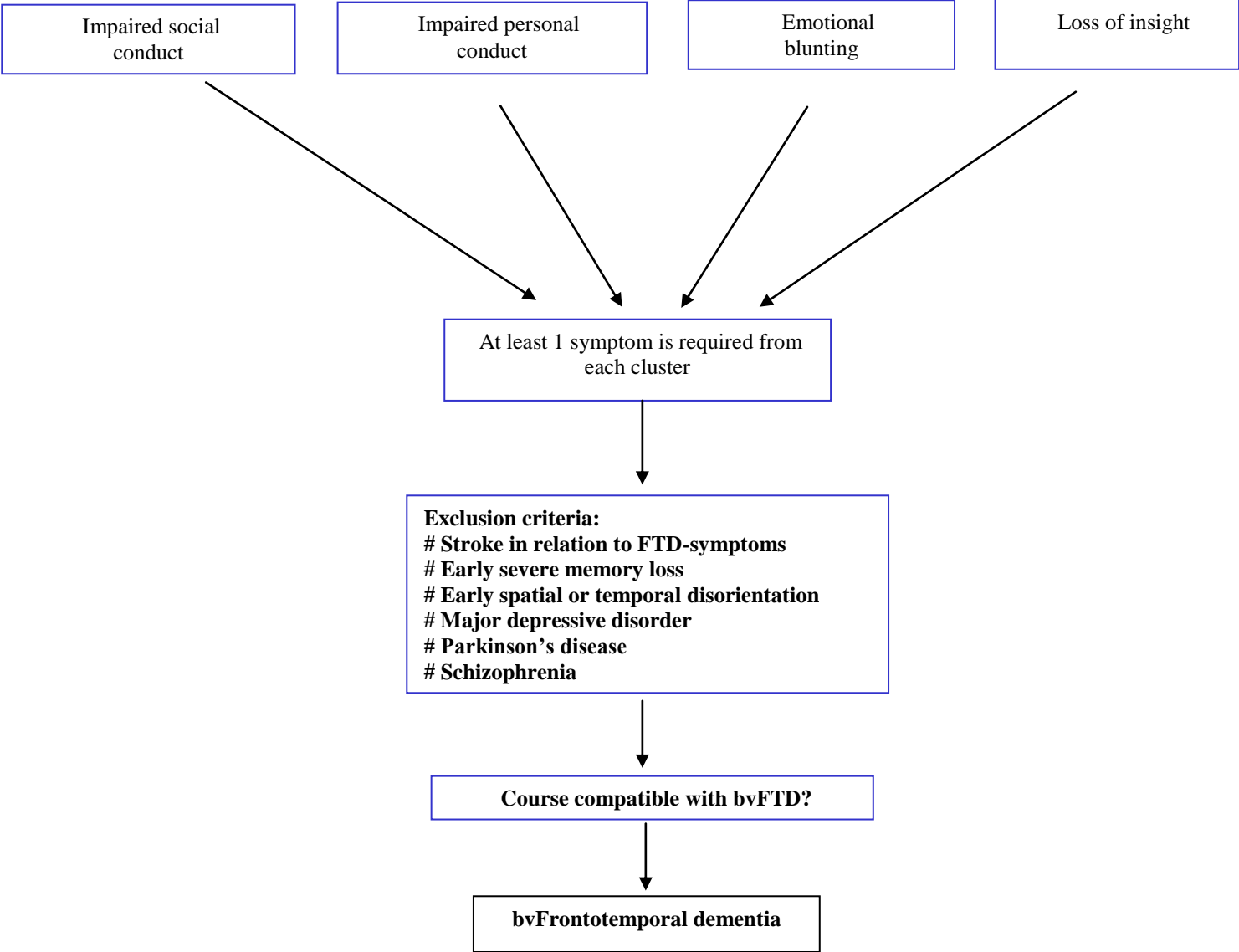


Figure 3. Algorithm for diagnosis of bvFTD from Lund-Manchester Research Criteria (LMRC).

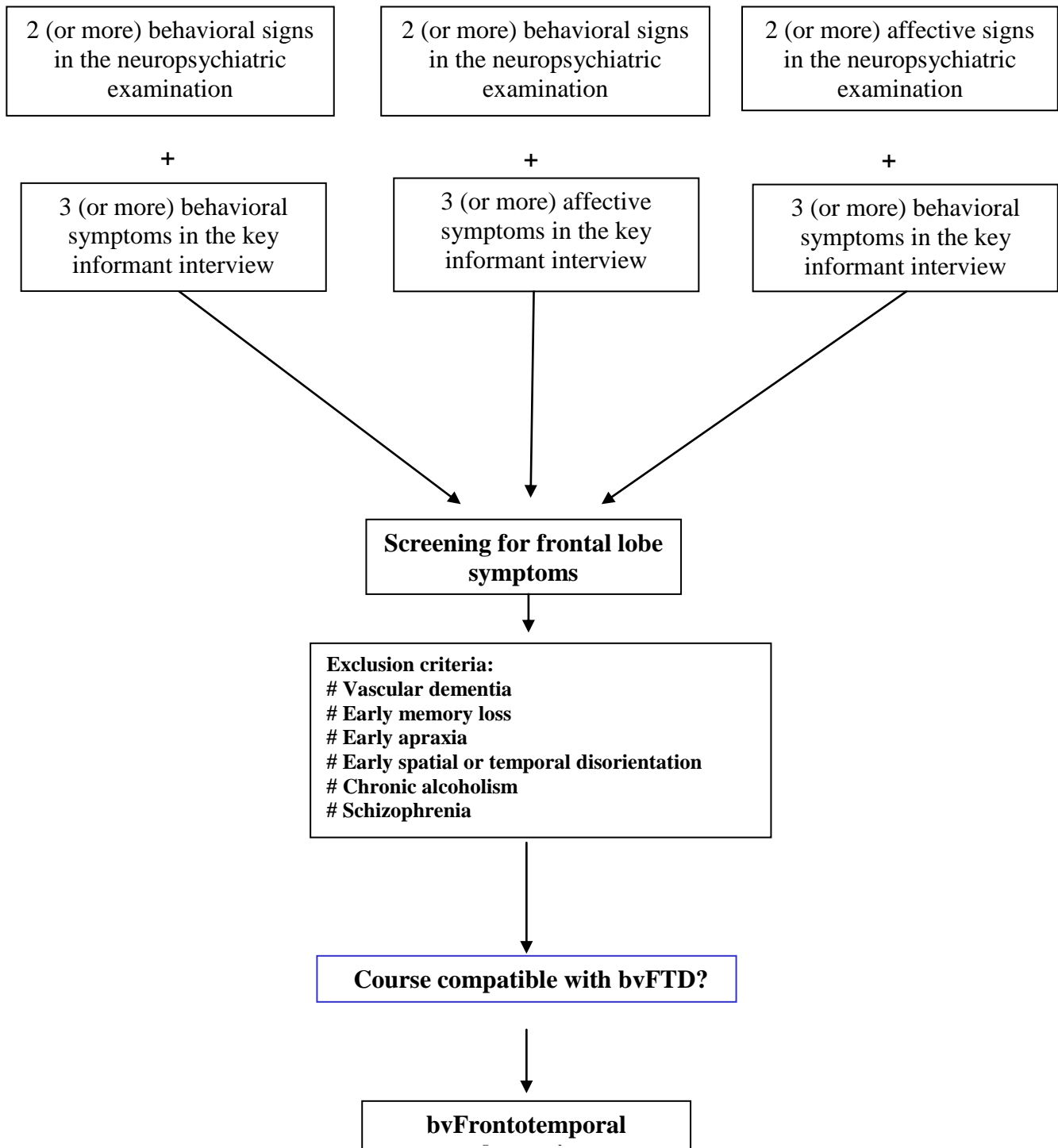


Fig. 4. Agreement between the FTDC, FTLD-CC and LMRC for behavioral variant frontotemporal dementia (bvFTD; total n=88).

